Isolated Aortic Stenosis in the Neonate
Natural History and Hemodynamic Considerations

By Jeffrey B. Lakier, M.D., Alan B. Lewis, M.D., Michael A. Heymann, M.D., Paul Stanger, M.D., Julien I. E. Hoffman, M.D., and Abraham M. Rudolph, M.D.

SUMMARY
We have reviewed the clinical and catheterization data and pathologic findings in ten infants under one month of age with isolated severe aortic valve stenosis. All presented with evidence of progressive cardiac failure and diminished cardiac output. A hyperdynamic right ventricular impulse was present in nine infants who were subsequently shown to have a left-to-right atrial shunt, through a stretched patent foramen ovale. Calculated aortic valve area ranged from 0.12 cm²/m² to 0.29 cm²/m². Three patients died prior to surgical intervention because of low cardiac output and refractory metabolic acidosis. Aortic valvotomy was attempted in the remaining seven. Five died intraoperatively or in the immediate postoperative period and one other died six months after surgery. Necropsy was obtained in all nine patients who died.

We believe that, in this group, the clinical course reflects fetal and postnatal hemodynamics. We have discussed the probable hemodynamic factors which occur and have related them to the morphological development of aortic stenosis in these infants. The poor prognosis of these patients probably results from a combination of factors which include left ventricular outflow obstruction, often associated with a small aortic annulus, a markedly thickened left ventricle, and left ventricular malfunction secondary to subendocardial ischemia and/or endocardial fibroelastosis.

Additional Indexing Words:
Myocardial ischemia
Cardiac catheterization
Congenital heart disease
Surgery

AORTIC STENOSIS is one of the more common congenital cardiac anomalies, occurring in 6% of infants with congenital heart disease;¹ the spectrum of severity ranges from critical aortic stenosis to a non-obstructive bicuspid valve. Although the natural history of aortic stenosis in children and adults has been reviewed,¹,² little attention has been directed to the natural history of severe aortic stenosis in the immediate newborn period. The one report in the literature which deals specifically with left ventricular outflow obstruction in this age group does not differentiate between isolated aortic stenosis and that associated with other complex cardiac anomalies.³ In infants under one month of age, severe aortic stenosis still is associated with a high mortality, despite advances in surgical technique.⁴,⁵

We have considered the possibility that this group of infants represents a different spectrum of the disease as compared with that occurring in older children. In infants with severe aortic stenosis the features of left ventricular development may be different compared with those of severe aortic stenosis in later life, and aortic stenosis in these infants may be more closely related to aortic atresia.

Ten cases of severe aortic stenosis presenting in infants under one month of age were reviewed. The hemodynamic and pathological findings in these infants were examined and related to possible prenatal circulatory dynamics and the changes in the circulation after birth. As other left-sided obstructive lesions such as severe aortic coarctation and aortic or mitral atresia may present clinical features similar to those of aortic stenosis in neonates, the necessity for documentation of the underlying cardiac abnormality at cardiac catheterization is stressed.

Methods
Ten patients aged one to 24 days were diagnosed at cardiac catheterization as having severe valvar aortic stenosis and in all the diagnosis was confirmed at operation and/or autopsy. Patients with mitral atresia, aortic atresia,

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hypoplastic or interrupted aortic arch, and aortic coarctation were excluded from the present study, as were those with aortic stenosis as part of other complex cardiac anomalies. The clinical, electrocardiographic, and roentgenographic features of the ten patients were analyzed. Blood gases were measured before and during cardiac catheterization and any base deficit corrected by parenteral administration of 0.9 M sodium bicarbonate. None of the infants had hypoglycemia or hypocalcemia. Cardiac catheterization was performed through a cutdown in the right groin. Arterial catheterization was performed through an umbilical artery in three infants and the superficial femoral artery in the other seven. The left ventricle was entered either passing the venous catheter across a patent foramen ovale and the mitral valve or by passing the arterial catheter across the aortic valve. The left ventricular-to-aortic systolic pressure difference was measured simultaneously (nine patients) or on withdrawal of the catheter across the aortic valve (one patient). Cardiac output was determined by the Fick method using an assumed value for oxygen consumption. Aortic valve area was calculated in seven patients using the modification of the Gorlin formula described by Bache et al. In comparative analyses we have shown that this estimation has similar values for aortic valve area when compared with the Gorlin formula. In order to assess an index of myocardial oxygen supply and demand, the ratio of the diastolic pressure time index (DPTI) multiplied by the arterial oxygen content to the systolic pressure time index (SPTI) was determined (DPTI × C/SPTI). The data on this ratio in four of the patients have also been included in a report by Lewis et al.

Left ventricular cineangiograms and aortograms were performed in nine of the ten patients. In one, only an aortogram was performed due to recurrent supraventricular tachycardia induced when the catheter was manipulated in the left ventricle. As normally administered volumes of contrast material carry a significant risk of inducing renal insufficiency in the presence of a reduced cardiac output, the number of angiograms at cardiac catheterization was kept to the minimum required to define the anatomy.

Three patients died before surgery could be performed. Operation was performed in seven infants and the anatomy of the aortic valve and aortic valve ring was assessed in each at operation. Five died intraoperatively or in the immediate postoperative period, and one six months postoperatively. Altogether there were nine deaths and necropsy was obtained in each case. One patient is alive and well 12 months postoperatively.

Results

The sex, age, and pertinent clinical findings at the time of cardiac catheterization in the ten infants are listed in table 1. There were three female and seven male patients. Central cyanosis was present twice (#8 and 10). All had evidence of respiratory distress with respiratory rates of from 60 to 120 per minute. Poor peripheral perfusion as evidenced by cool extremities, duskeness or peripheral cyanosis and arterial pulses of poor volume (carotids, axillary and femoral) were prominent features. A progressive decrease in pulse volume, over 12 hours to 3 days, was noted in three patients while under observation. Hepatomegaly was present in all. A hyperactive right ventricular impulse over the precordium was present in nine patients and progressively increasing hyperactivity was noted in four during the 12 to 72 hours prior to cardiac catheterization. None of the patients had a dominant left ventricular impulse.

Table 1

Relevant Clinical Data on Ten Neonates with Aortic Stenosis

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (days)</th>
<th>Sex</th>
<th>Systolic ejection clicks</th>
<th>Systolic murmur grades 1-6</th>
<th>R.V. impulse</th>
<th>Electrocardiogram</th>
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<tbody>
<tr>
<td></td>
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<td></td>
<td></td>
<td></td>
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<td>F</td>
<td>+</td>
<td>3→→0</td>
<td>+→→+</td>
<td>+110°</td>
</tr>
<tr>
<td>2</td>
<td>21</td>
<td>M</td>
<td>+</td>
<td>3</td>
<td>+</td>
<td>+110°</td>
</tr>
<tr>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>5</td>
<td>M</td>
<td>+</td>
<td>3→→1</td>
<td>+→→+</td>
<td>0°</td>
</tr>
<tr>
<td>4</td>
<td>24</td>
<td>M</td>
<td>-</td>
<td>2</td>
<td>-</td>
<td>+110°</td>
</tr>
<tr>
<td>5</td>
<td>20</td>
<td>M</td>
<td>+</td>
<td>3</td>
<td>+</td>
<td>+70°</td>
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<td>6</td>
<td>11</td>
<td>F</td>
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<td>F</td>
<td>+</td>
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<td>8</td>
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<td>M</td>
<td>-</td>
<td>2</td>
<td>+</td>
<td>+130°</td>
</tr>
<tr>
<td>9</td>
<td>2</td>
<td>M</td>
<td>-</td>
<td>3</td>
<td>+</td>
<td>+140°</td>
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<tr>
<td>10</td>
<td>1</td>
<td>M</td>
<td>-</td>
<td>3</td>
<td>+</td>
<td>+120°</td>
</tr>
</tbody>
</table>

Abbreviations: RV = right ventricle; RVH = right ventricular hypertrophy; LVH = left ventricular hypertrophy.
On auscultation the first heart sound was of normal intensity. An ejection systolic click was heard in six patients, in one of whom the ejection click was heard only at the initial examination and not subsequently. A grade 2–3/6 ejection systolic murmur which occupied about the first two-thirds of systole was present in all patients when first examined but in three (‡3, 6 and 7) the murmur was noted to decrease in intensity and it became inaudible in a fourth patient (‡1). The decreasing intensity of the murmur was associated with increasing precordial hyperactivity and hepatomegaly and a decrease in peripheral perfusion in these four patients. A prominent gallop rhythm was present at the apex in eight patients. The lung fields revealed rales in only one patient and generalized wheezing in another. The remaining patients had clinically clear lung fields.

The electrocardiographic findings were variable. The mean frontal plane QRS axis varied from 0° to 140°. Six patients had evidence of right ventricular hypertrophy (fig. 1), three had left ventricular hypertrophy (fig. 2), and one had an electrocardiogram that was within normal limits for age. Absent or diminished left ventricular forces were present in three (‡2, 9 and 10). Inverted T waves and depressed ST segments were present across the left chest leads or inferior limb leads in seven patients. Suggestive evidence of right atrial enlargement was present in three patients and left atrial enlargement in four.

Chest roentgenograms demonstrated an increased cardiothoracic ratio in all ten infants. Lung vascularity was normal or increased, but pulmonary venous congestion was thought to be present on only two of the roentgenograms.

The information obtained at cardiac catheterization is listed in table 2. A systolic pressure drop across the aortic valve ranging from 20 to 120 mm Hg was present in all patients. Mean aortic pressure was normal, but pulse pressure was diminished (range 5 to 24 mm Hg). Left ventricular systolic pressure ranged from 90 mm Hg to 170 mm Hg and left ventricular end-diastolic pressures were elevated to 15 to 37 mm Hg. Left-to-right shunting at the atrial level, as evidenced by significant increases in oxygen saturations in the right atrium, was present in nine patients; the shunt was confirmed at cineangiography in seven. Three of these patients (‡8, 9, 10) also had right-to-left ductus shunting as evidenced by a decrease in oxygen saturation in the descending aorta as compared with the ascending aorta and left ventricle. In these three patients the ductus right-to-left shunt precluded meaningful calculation of systemic output by the Fick method. In four of the remaining seven patients the calculated cardiac index was below 3.0 liters/min/m². In all seven patients in whom an aortic valve area could be calculated it was below 0.29 cm²/m² (range 0.12 cm²/m² to 0.29 cm²/m²) and was thus well below the figure of 0.7 cm²/m² considered to indicate severe aortic stenosis.10, 11 All patients had DPTI/SPTI ratios below 0.7 and DPTI × C/SPTI ratios below 10.0.
Left ventricular cineangiograms were obtained in nine patients. The left ventricular wall was considerably thickened irrespective of the size of the left ventricular cavity. Left ventricular cavity size was normal or slightly increased in eight patients, but was reduced in two (#9 and 10). The aortic valve was thickened and domed during systole. The ascending aorta was dilated in five infants, and was of normal dimension in the other five infants. Five patients had mild and two had moderate mitral regurgitation. One patient had a small ductus left-to-right shunt detected on an aortogram. Three patients redeveloped persistent, severe metabolic acidosis after cardiac catheterization and died prior to operative intervention despite vigorous medical therapy.

Aortic valvotomy was attempted in seven infants. Redundant aortic valve tissue was also excised in one of these. Five died intraoperatively or in the immediate postoperative period because of inadequate cardiac output. Two infants survived surgery; case #1 had a functional bicuspid aortic valve with a diminutive right coronary cusp. One year later she has a grade 3/6 aortic ejection systolic murmur lasting 2/3 of systole and a grade 2/6 aortic early diastolic murmur. She is on maintenance digoxin and a diuretic, but is developing normally. Case #3 died six months postoperatively, after a course complicated by repeated episodes of congestive cardiac failure despite digitalis and diuretic therapy.

Necropsy data was obtained in the nine patients who died (table 3). In all, the dominant features were left ventricular hypertrophy and thickening of the stenotic aortic valve. The anatomy of the aortic valve was variable; it was tricuspid in three, bicuspid in four, and unicusp in two. In all the mitral valve was normally formed but the mitral valve ring was smaller than normal in cases #9 and 10. Seven patients had a small aortic valve annulus. Seven of the nine patients had a normal or dilated left ventricular cavity. Two of the three youngest patients (#9 and 10) had small left ventricular cavities, and all three (#8, 9, 10) had a patent ductus arteriosus and had been noted to have right-to-left ductus shunting at cardiac catheterization. Surgical or necropsy information regarding the interatrial communication was available in eight patients within one week of cardiac catheterization. A large patent foramen ovale was closed at surgery in case #2. Seven patients (#4-10) had at necropsy a patent foramen ovale, which was measured in four infants and was found to be between 5 and 10 mm in diameter. Case #10 had a probe patent foramen ovale, but in addition had left-to-right shunting at cineangiography through a levo-atrial cardinal vein which communicated with the left innominate vein and drained into the right atrium.

The three youngest patients (#8, 9, 10) and two others (#5, 6) had evidence of endocardial fibroelastosis of the left ventricle both on gross inspection and histologically.

Discussion

The ten infants presented in this report represent an intermediate group between infants with aortic atresia and hypoplastic left heart syndrome on the one hand and aortic stenosis occurring later in life on the other. As opposed to patients with hypoplastic left heart syndrome, the ascending aorta in all our patients was of either normal dimensions or dilated. In only two in-
We reported by

In left-sided lesion (cases after birth, Circulation, Volume and postnatal (150 output. Without this that normally ventricular about descending foramen crosses ml/kg/min (44% of is from inferior 26%.

Premature closure of the foramen ovale is conceivable that with lesser degrees of obstruction to the foramen ovale, less severe left heart underdevelopment could result, with a concomitant wide range of left ventricular size. Hypoplastic left heart syndrome has also been experimentally produced in the chick embryo by interference with normal development of the left atrioventricular canal. Varying degrees of left heart hypoplasia, including a stenotic thickened aortic valve and hypoplastic ascending aorta, resulted and were attributed to differences in the amount of flow entering the left ventricle. Either of these two mechanisms might have been operative in cases #9 and 10.

Primary involvement of the aortic valve during fetal life could, however, also explain the morphological findings. The severity of the aortic valve obstruction and the gestational period at which it develops could influence the development of the left ventricle. In the fetal lamb 0.6-0.7 (90-105 days) gestation, moderately severe supravalvar obstruction, produced by constriction of the aorta, resulted in a marked decrease in left ventricular output associated with a rapid increase in left ventricular wall thickness. Mean left ventricular volumes were less than half those of matched controls in these relatively acute preparations, followed for about three weeks. The increase in muscle mass developed in response to the obstruction could result in a decrease in left ventricular compliance, thus interfering with left ventricular filling. Left atrial and left ventricular end-diastolic pressure would not increase significantly, but rather flow into the left atrium across the foramen ovale would decrease.

On the basis of these findings we suggest that severe aortic obstruction produced very early in gestation would result in thickening of the left ventricular wall with a marked reduction in cavity size: this could result in a true hypoplastic left ventricle. Cases #9 and 10 in whom left ventricular size was diminished could be explained on this basis. If the stenosis was less severe or occurred later in gestation the left ventricular wall would still be thickened, but left ventricular cavity size would not be significantly reduced.

Normally after birth, with elimination of the placental circulation, the decrease in pulmonary

<table>
<thead>
<tr>
<th>Case number</th>
<th>Left ventricle (S)</th>
<th>Right ventricle (D)</th>
<th>Aorta (S)</th>
<th>Aortic valve area (cm²/m²)</th>
<th>DPTI/SPTI</th>
<th>DPTI × C/SPTI</th>
<th>Qp:Qs</th>
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<td>110</td>
<td>36</td>
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<td>45</td>
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<td>4.0</td>
<td>0.28</td>
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<td>38</td>
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<td>42</td>
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</table>

Abbreviations: DPTI = diastolic pressure time index; SPTI = systolic pressure time index; C = oxygen content (ml/100 ml); Qp:Qs = pulmonary/systemic flow ratio; S = systolic; D = end-diastolic; M = mean.
### Table 3

Pathology Data on Nine Neonates with Congenital Isolated Valvar Aortic Stenosis

<table>
<thead>
<tr>
<th>Case no.</th>
<th>RV</th>
<th>LV</th>
<th>Aortic valve</th>
<th>Ascending aorta</th>
<th>Inter-atrial defect</th>
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<tr>
<td></td>
<td>Cavity size</td>
<td>Wall thickness (mm)</td>
<td>Cavity size</td>
<td>Wall thickness (mm)</td>
<td>LA size</td>
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<tr>
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<td>Dilated</td>
<td>5</td>
<td>Normal</td>
<td>8</td>
<td>Normal</td>
</tr>
<tr>
<td>3</td>
<td>Dilated &amp; hypertrophied Normal</td>
<td>Normal</td>
<td>Thick walled</td>
<td>Dilated</td>
<td>5.0</td>
</tr>
<tr>
<td>4</td>
<td>Slightly dilated Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>8</td>
<td>Normal</td>
</tr>
<tr>
<td>5</td>
<td>Dilated</td>
<td>4</td>
<td>Slightly dilated</td>
<td>10</td>
<td>3.0–4.0</td>
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<td>6</td>
<td>Slightly dilated Normal</td>
<td>11</td>
<td>Dilated</td>
<td>Normal</td>
<td>Unicuspid, rolled &amp; thickened</td>
</tr>
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<td>7</td>
<td>Normal</td>
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<td>6</td>
<td>6.0</td>
<td>Bicuspid, polypoid &amp; slightly translucent</td>
</tr>
<tr>
<td>8</td>
<td>Slightly dilated Normal</td>
<td>10</td>
<td>Normal</td>
<td>Small</td>
<td>Bicuspid, thickened valve</td>
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<td>Small</td>
<td>6</td>
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<td>Small</td>
<td>Small</td>
</tr>
<tr>
<td>10</td>
<td>Dilated</td>
<td>–</td>
<td>Small</td>
<td>Thick walled</td>
<td>Small</td>
</tr>
</tbody>
</table>

Abbreviations: PFO = patent foramen ovale; LACV = Levo-atrial cardinal vein; RV = right ventricle; LA = left atrium; LV = left ventricle.
vascular resistance and closure of the ductus arteriosus, total right ventricular output which amounts to about 200 ml/kg/min is ejected into the pulmonary circulation. The total pulmonary flow returns to the left atrium and left ventricle with a consequent increase of left ventricular output from 150 ml/kg/min prenatally to 200 ml/kg/min postnatally, an increase of about 25%.

Postnatally, the survival of patients with severe aortic stenosis depends on the ability of the left ventricle to maintain systemic flow. If the left ventricle cannot do so, the ductus may perform an important function in maintaining systemic output. The three youngest patients, aged 18 hours to 2 days, had evidence at cardiac catheterization of a patent ductus arteriosus. All three were in marked congestive cardiac failure and two of them had small left ventricular cavities demonstrated cineangiographically and at necropsy. Descending aortic flow in these patients was probably dependent on the right-to-left ductal shunt. Severe persistent metabolic acidosis, despite the administration of sodium bicarbonate, was present following cardiac catheterization and all three patients died before surgery. It is probable that progressive ductal closure in these infants resulted in diminishing aortic flow and organ perfusion. The clinical course of these infants was therefore similar to that of infants with aortic atresia or hypoplastic left heart syndrome.

In the remaining seven patients, although a cardiac murmur was heard on the first postnatal day, myocardial failure only became evident three days to three weeks after birth. Continued patency of the ductus arteriosus might have played a role in delaying the onset of symptoms in these infants; however, by the time of study the ductus arteriosus was functionally closed in all seven patients. Under these circumstances right ventricular output must return via the pulmonary circulation to the left atrium as in the normal. With severe aortic stenosis the 25% increase in flow returning to the left ventricle would impose a significant volume overload on the hypertrophied left ventricle and is reflected by the raised left ventricular end-diastolic pressure (range 15-36 mm Hg) found at cardiac catheterization.

In nine patients (#1-3, 5-10) there was a hyperactive precordial right ventricular impulse in association with poor peripheral perfusion. A striking clinical feature in four patients was that precordial activity increased as peripheral perfusion decreased. In three of these latter patients the systolic ejection murmur decreased in intensity. At cardiac catheterization all nine had a significant increase in oxygen saturation in the right atrium, indicative of left-to-right atrial shunting. Postnatally, as a result of the normal pulmonary venous return to the left atrium and the presence of a thickened left ventricle, left atrial pressures are elevated. This may result in stretching of the foramen ovale which could explain the finding of an enlarged foramen ovale in four patients and herniation of the valve into the right atrium in one of these. Should this occur, left-to-right atrial shunting postnatally would be encouraged. The result might be a decrease in left ventricular filling pressure and a decrease in left ventricular output and could account for the decreasing intensity of the systolic murmur. The left-to-right atrial shunt would result in volume overload of the right ventricle and would explain the clinical finding of right ventricular hyperactivity. Increasing left ventricular end-diastolic pressure would result in a progressive increase in the left-to-right atrial shunt and right ventricular hyperactivity, with a concomitant decrease in systemic output. The frequently observed clinical and electrocardiographic evidence of right ventricular hypertrophy in these infants could thus result from a possible increase of right ventricular output in the fetus or from left-to-right atrial shunting postnatally or from a combination of both mechanisms.

In view of the high incidence of subendocardial fibrosis seen in this group of infants, we have reviewed the DPTI/SPTI ratio.18 The combination of an increased left ventricular systolic pressure relative to aortic pressure and raised left ventricular end-diastolic pressure seen in our patients should result in a marked decrease of subendocardial coronary flow. This is borne out by DPTI/SPTI ratios which in all cases was considerably below the figure of 0.7 found to be associated with experimental subendocardial ischemia.18 A more accurate estimation of oxygen delivery to the myocardium is obtained if DPTI/SPTI is multiplied by aortic oxygen content.8,9 Subendocardial ischemia in dogs has been shown to be present when this ratio is reduced below 10. Once again all our patients had ratios in the range associated with ischemia. The occurrence of subendocardial fibrotic changes and the association of endocardial fibroelastosis in this group of infants may thus be related to impaired subendocardial oxygen delivery.1,17

The poor long-term results in this group of patients are disappointing and dramatic improvement following surgery is uncommon. The exact reasons for this are not known. Commisurotomy may not ensure an effective aortic valve orifice to allow adequate aortic blood flow without producing significant aortic regurgitation. Furthermore, the myocardium, malfunctioning both as a result of subendocardial ischemia and endocardial fibroelastosis, and markedly hypertrophied with resultant decreased compliance, may be unable to maintain a normal output. Since a proportion of infants, less than 1 month of age, do
well, at least in the immediate postoperative period.\(^4\)\(^,\)\(^6\) an aggressive approach is necessary in all infants with suspected severe valvar aortic stenosis. Awareness of the condition with earlier detection may result in improved survival. Careful attention to intraoperative and postoperative care with particular reference to myocardial perfusion may also improve the outcome. Other conditions, notably aortic coarctation, which has a far better prognosis and is eminently correctable, may present clinically in a similar manner. Cardiac catheterization therefore remains essential to confirm the diagnosis.

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