Subaortic stenosis, the univentricular heart, and banding of the pulmonary artery: an analysis of the courses of 43 patients with univentricular heart palliated by pulmonary artery banding

ROBERT M. FREEDOM, M.D., F.R.C.P.(C), F.A.C.C., LEE N. BENSON, M.D., F.R.C.P.(C), JEFFREY F. SMALLHORN, M.D., F.R.C.P.(C), WILLIAM G. WILLIAMS, M.D., F.R.C.S.(C), GEORGE A. TRUSLER, M.D., F.R.C.S.(C), AND RICHARD D. ROWE, M.D., F.R.C.P.(C)

ABSTRACT  Subaortic stenosis is well known to complicate the clinical course of patients with single ventricle or univentricular hearts, and we have previously suggested that the development of subaortic stenosis in such patients may be causal to and/or accelerated by previous banding of the main pulmonary trunk. To further define the relationship between banding of the pulmonary artery in patients with univentricular hearts and the development of subaortic stenosis, we examined the morphologic substrate and timing of the development of subaortic stenosis in 43 patients seen at our institution from January 1, 1970, through June 30, 1985. These 43 patients include all patients in this period with an unequivocal univentricular heart whose longitudinal data was available for follow-up. We excluded patients who died within 1 week of surgery, patients lost to follow-up, and patients with evidence of subaortic stenosis before banding. Thirty-one of 43 patients (72.1%) developed subaortic stenosis subsequent to banding of the main pulmonary artery. The mean age at banding of those patients who developed subaortic stenosis was 0.21 years, and subaortic stenosis was recognized at a mean age of 2.52 years. For the specific cohort of patients whose ventricular morphology was a main chamber of left ventricular type supporting the pulmonary artery and a rudimentary right ventricle supporting the transposed aorta (32 patients), 27 developed subaortic stenosis (84.4%). Subaortic stenosis in the classic form of single ventricle usually results from progressive restriction of a wholly muscular interventricular communication. Banding of the pulmonary artery by producing myocardial hypertrophy undoubtedly accelerates the potential for subaortic stenosis in these patients. Furthermore, one must realize that subaortic stenosis may be present in the absence of a resting pressure gradient, and such subaortic stenosis can usually be unmasked by stimulation with isoprenaline. Finally, one must be guarded in advocating banding of the pulmonary artery in patients with single ventricle, realizing that subaortic stenosis strongly influences the outcome of more definitive surgery in these patients.

Circulation 73, No. 4, 758–764, 1986.

THE NATURAL HISTORY of patients with single ventricle (one-ventricle hearts, univentricular hearts, univentricular atrioventricular connection) has been irrevocably altered by those diverse surgical procedures that (1) augment pulmonary blood flow, (2) reduce pulmonary blood flow, (3) augment atrial mixing when intracardiac streaming is disadvantageous, or (4) separate the systemic venous from the pulmonary venous circulation.1–4 Ideally, any preliminary surgical procedure should protect the integrity both of the pulmonary vascular bed and the myocardium, thus setting the stage for later more definitive long-term anatomic palliation.

From a variety of clinical reports, it is clear that subaortic stenosis may complicate the course of patients with univentricular heart.5–14 The presence of subaortic stenosis is certainly an incremental risk factor in patients subjected either to ventricular septation or to an atrio pulmonary anastomosis of the Fontan type.12–17 We have previously suggested that the recognition of subaortic stenosis in the patient with a univen-
tricular heart usually implies a discordant ventriculoarterial connection (or transposition of the great arteries) and that the documentation of subaortic stenosis usually follows pulmonary artery banding. Furthermore, we have stated that pulmonary artery banding may be causal to the genesis of subaortic stenosis or at the very least may accelerate its development.

To define the "unnatural" history of patients with a univentricular heart who have undergone palliative pulmonary artery banding, we have examined the morphologic substrate and clinical course of 43 such patients seen at The Hospital for Sick Children, Toronto, between January 1, 1970, and June 30, 1985. These patients were examined to determine the incidence of subaortic stenosis, the timing of its recognition, and the number of patients in the initial cohort who underwent successful ventricular septation, atrioventricular surgery of the Fontan type, or arterial repair with construction of an atrioventricular connection.

Materials and methods

Ventricular morphology. There is not unanimity about the nomenclature concerning those hearts variously described as one-ventricle hearts, single ventricle, univentricular hearts, or hearts exhibiting a univentricular atrioventricular connection. For the purpose of this study, we have arbitrarily confined our analysis to those hearts with (1) a main left ventricle, a rudimentary right ventricle or outlet chamber, with the rudimentary chamber receiving none or tenuous atrioventricular valve connections (figure 1) — the atrioventricular connections in this and the other forms of univentricular hearts include double or common inlet and absent right or left atrioventricular connection; (2) a single ventricle that is apparently morphologically right without a hypoplastic morphologically left ventricle; and (3) a ventricle whose morphology is neither clearly morphologically right nor left, which is considered of indeterminate morphology.

Diagnosis of subaortic stenosis in the univentricular heart. We and others have shown elsewhere that in the clinically stable patient, the absence of a resting pressure gradient between the single ventricle and aorta does not exclude subaortic stenosis. We consider any resting pressure gradient between the left ventricle and aorta as evidence for subaortic stenosis, although others adhere to a minimal resting gradient (figure 2). In the absence of a resting pressure gradient, we share both the view and experience of Somerville et al.  that a pressure gradient provoked by isoprenaline is indicative of subaortic stenosis.

Morphologic and clinical observations have also shown that among some newborn babies and neonates dying with this type of congenital heart disease, the ventricular septal defect can be restrictive, resulting in subaortic stenosis. Such babies are usually terribly ill, with severe congestive heart failure, tremendous pulmonary blood flow, pulmonary artery hypertension at systemic levels, and a low cardiac output. An obstructive anomaly of the aortic arch may be present (figure 3). One is not surprised, then, that a pressure gradient between aorta and single ventricle may not be recorded even when the ventricular septal defect is obviously restrictive. Thus the preoperative management of such babies mandates that attention be focused on the size of the ventricular septal defect relative to aortic root and that the defect be scrutinized for the reality or potential of subaortic stenosis. Such scrutiny may include Doppler echocardiographic, angiocardiographic, or pharmacologic (i.e., isoproterenol challenge) investigations.

Patients. All patients were seen at The Hospital for Sick Children, Toronto, between January 1, 1970, and June 30, 1985. Cardiac records were searched from a computer printout for the diagnosis of univentricular heart (see Definitions), and a complete surgical logbook of all patients (with cardiac anatomic diagnoses) subjected to banding of the main pulmonary trunk was scrutinized and compared individually with our cardiac records. In addition, a complete logbook of all angiograms performed from January 1, 1970, through June 30, 1985, was examined and patients with the diagnosis of univentricular heart, single or common ventricle, or tricuspid and mitral atresia were retrieved; the charts were examined and compared with the surgical list. These maneuvers ensured a complete patient listing.

Excluded from the analysis was any patient with a univentricular heart who had unequivocal evidence of subaortic stenosis before palliative pulmonary artery banding. Furthermore, we did not enter into the data base any patient in whom the diagnosis of univentricular heart was equivocal (i.e., a biventricular heart with straddling of an atrioventricular valve, etc.). We also arbitrarily excluded patients who died within 1 week of the banding procedure, and patients who underwent banding at this institution but were lost to follow-up (two patients).

Forty-three patients were identified as having both a single ventricle (univentricular heart) and banding of the main pulmonary artery. One patient had undergone banding elsewhere in early infancy, but his long follow-up at our institution and the ability to review his prebanding hemodynamic and angiographic data prompted us to include him. These 43 patients included

FIGURE 1. Large ventricular septal defect or outlet foramen (asterisks) in a patient with previously banded pulmonary artery (PA), absent left atrioventricular connection, univentricular heart of left ventricular type, rudimentary right ventricle (RV), and transposition of the great arteries. This elongated left axial oblique right ventriculogram profiles the very large interventricular communication and the infundibular septum (IS). It would be unlikely for such a large defect to become smaller.
FIGURE 2. Angiograms from a 7-year-old child with absent left atrioventricular connection, univentricular heart of the left ventricular type, left-sided rudimentary right ventricle, transposition of the great arteries, and banded pulmonary trunk. Left, This retrograde angiogram shows a hypoplastic infundibular chamber (rv) supporting the aorta. Right, The interventricular communication (asterisk) is obviously restrictive (75 mm Hg gradient).

FIGURE 3. Angiograms from a baby found to have a restrictive ventricular septal defect 1 month after coarctectomy and banding of the main pulmonary trunk. Left, Frontal left ventricular angiogram shows a restrictive defect (asterisk). Right, This diastolic frame shows the superiorly positioned rudimentary right ventricle (rv) and aorta (ao). It is conceivable that banding of the pulmonary trunk may have unmasked the restrictive ventricular septal defect (by changing a volume-loaded ventricle to a pressure-loaded one). LV = left ventricle.
all those with univentricular hearts subjected to palliative banding (with the exclusions noted above) from January 1, 1970, through June 30, 1985, whose follow-up data were available for review. Finally, certain aspects of these 43 patients have been included in previous publications from this institution.

Results

Twenty-nine patients were boys and the remainder were girls. The basic anatomic types of univentricular heart among these 43 patients are shown in table 1.

Age at banding of the pulmonary trunk. All patients had undergone banding of the main pulmonary artery. The age at banding for the entire group ranged from less than 1 day to 3 years (mean 0.23 years).

Subaortic stenosis. Subaortic stenosis was diagnosed in 31 patients (72%). These patients ranged in age at the time of banding from less than 1 day to 1.07 years (mean 0.21 years). Their ages at the time subaortic stenosis was unequivocally documented (during life) ranged from 0.05 to 8.07 years (mean 2.52). The morphologic substrates and frequency of subaortic stenosis are shown in table 1.

Diagnosis of subaortic stenosis. A resting pressure gradient ranging from 0 to 105 mm Hg (mean 55.6) was recorded in 20 patients at cardiac catheterization. In the absence of a resting pressure gradient, isoprenaline provoked a gradient in seven patients ranging from 30 to 110 mm Hg. The diagnosis of subaortic stenosis was made only at autopsy in four patients.

Surgical results. We have previously reviewed our surgical experience with many of these patients. As of June 30, 1985, of the 31 patients who developed subaortic stenosis subsequent to pulmonary artery banding, nine patients are alive. Two patients are doing very well after undergoing both an arterial switch and atrioventricular connection of the Fontan type. Three patients have undergone palliation by surgical enlargement of the restrictive ventricular septal defect, and another two patients have undergone palliation by surgical enlargement of the ventricular defect in combination with an insertion of a left ventricular–aortic conduit. Another patient has had palliation by construction of an aortopulmonary window proximal to the band and is currently awaiting further surgery. For one patient, the parents have refused surgical intervention to enlarge a restrictive interventricular communication. We have no long-term survivors of either aortopulmonary anastomosis with connection of the proximal pulmonary artery to the ascending aorta nor of ventricular septation.

Discussion

Banding of the main pulmonary artery continues to be advocated in the palliation of patients with univentricular heart, torrential pulmonary blood flow, and pulmonary artery hypertension with congestive heart failure. Data obtained from 5 years experience in the New England Regional Infant Cardiac Program on the results of pulmonary artery banding showed that only 22 patients (20 with single ventricle, two with tricuspid atresia) from a total 1916 infants with congenital heart disease registered into the survey underwent banding of the main pulmonary trunk. Of these 22 patients, 12 survived to 1 year of age and were theoretically considered as correctable. There are few data, however, on

TABLE 1

Types of univentricular hearts included in this analysis

<table>
<thead>
<tr>
<th>Type of heart</th>
<th>No.</th>
<th>No. with subaortic stenosis</th>
<th>Percent developing subaortic stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid atresia, VSD, unguarded pulmonary blood flow (so-called tricuspid atresia, type 1C)</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Single ventricle of right or indeterminate morphology</td>
<td>3</td>
<td>1</td>
<td>33.3</td>
</tr>
<tr>
<td>Morphologic left ventricle, rudimentary right ventricle, and normal ventriculoarterial connections</td>
<td>6</td>
<td>3</td>
<td>50</td>
</tr>
<tr>
<td>Double-inlet AV connection (5 patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent left AV connection (1 patient)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Morphologic left ventricle, rudimentary right ventricle, and discordant or transposed ventriculoarterial connection</td>
<td>32</td>
<td>27</td>
<td>84.4</td>
</tr>
<tr>
<td>Absent right AV connection (10 patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Double-inlet AV connection (19 patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent left AV connection (3 patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>43</td>
<td>31</td>
<td>72.1</td>
</tr>
</tbody>
</table>

VSD = ventricular septal defect; AV = atrioventricular.
the long-term results of banding of the main pulmonary artery in any cohort of patients with univentricular hearts specifically focusing on the development of subaortic stenosis.

We now present persuasive data that link the development of subaortic stenosis in the patient with univentricular heart to previous banding of the pulmonary artery. The data from our institution over the 15 year study period indicate that 84.4% of patients with the classic form of univentricular heart (i.e., major chamber having left ventricular morphology, rudimentary right ventricle, and transposition of the great arteries) subjected to pulmonary artery banding will develop subaortic stenosis. Data culled from the literature show that of 52 patients with this type of single ventricle, transposition of the great arteries, and subaortic stenosis, 44 have undergone pulmonary artery banding (84.6%).

Our own observations and those recorded in the literature clearly indicate that the ventricular septal communication is for the most part intrinsically larger in those patients with single ventricle, transposition of the great arteries, and pulmonary outflow tract obstruction than that in patients with unobstructed pulmonary blood flow. Indeed, when an obstructive anomaly of the aortic arch is also present, the ventricular septal defects tend to be equal to or smaller than the corresponding aortic root. Thus, in this latter group (those with unobstructed pulmonary blood flow), in which the interventricular communication tends not to be large, one might anticipate spontaneous diminution in the size of the defect.

Additionally, it should be emphasized that the interventricular communications in patients with the usual form of univentricular heart are not membranous. Rather, these defects are wholly muscular, and thus their dimensions are vulnerable to myocardial hypertrophy. Rather impressive myocardial hypertrophy can evolve quickly after banding of the pulmonary artery. From our extensive clinical data and from observations recorded in the literature, we suspect that in some young infants banding unmasks a restrictive ventricular septal defect (and thus subaortic stenosis), and by causing myocardial hypertrophy, it is both causal to and accelerates the process of subaortic stenosis in those patients with the appropriate morphologic substrate.

If the ultimate function of palliation of the patient with single ventricle is to preserve both the integrity of the pulmonary vascular bed and the myocardium, then in most patients with the usual form of univentricular heart, one would have to view pulmonary artery banding with some degree of skepticism. Our data indicate the majority of patients subjected to banding of the main pulmonary trunk will develop subaortic stenosis and (if untreated) profound myocardial hypertrophy. The histopathologic sequelae to obstruction to both arterial outlets is myocardial cellular hypertrophy and myocardial ischemia-fibrosis. Furthermore, there is an increasing body of data suggesting that banding of the main pulmonary artery may not protect the vascular bed from morphologic alterations that would jeopardize successful atrio pulmonary surgery, i.e., extension and increase in pulmonary vascular smooth muscle. Such pulmonary vascular abnormalities may be exaggerated in patients with left atrioventricular valve atresia or stenosis and restriction (even mild) at atrial level or with subaortic stenosis. Our data (previously published and unpublished) indicate that in the presence of significant subaortic stenosis, despite pulmonary artery banding, the distal pulmonary artery pressure may increase. Additionally, myocardial hypertrophy, by altering ventricular compliance, will impair atrial emptying, and this too may contribute to an altered pulmonary vascular bed.

What, then, are the implications of these observations? We are not suggesting that banding of the main pulmonary trunk be arbitrarily excluded as a form of palliation for patients with univentricular hearts. One must be cognizant, however, of the potential for subaortic stenosis and the consequences of its development. On the basis of such observations and clinical experiences in our institution, we have now altered our approach to the initial surgical management of neonates and young infants with actual or potential subaortic stenosis within the anatomic matrix of a univentricular heart. Rather than band the main pulmonary trunk and await the development of severe subaortic stenosis (and then dealing with it), we have taken two different surgical approaches in this difficult group of patients. Some neonates and young infants have undergone a Norwood type procedure, where the pulmonary trunk is transected, the proximal pulmonary artery is connected to the aorta, and a modified (4 to 5 mm Goretx) Blalock-Taussig shunt is connected to the distal pulmonary arteries. Another group of babies has undergone the palliation (interposition of a graft from the main pulmonary artery to the descending aorta, banding the proximal main pulmonary artery) advocated by Litwin et al. some years ago in the treatment of subaortic stenosis complicating ventricular septal defect and interruption of the aortic arch. Whether such procedures will provide a myocardial...
matrix and pulmonary vascular bed favorable for either atrial partitioning and atriopulmonary surgical connection or ventricular septation is as yet unknown.

References

35. Rahimtoola SH, Ongley PA, Swan HFC: The hemodynamics of common (or single) ventricle. Circulation 34: 14, 1966
45. Anderson RH, Penkoske PA, Zuberbuhler JR: Variable morphol-
ogy of ventricular septal defect in double inlet left ventricle. Am J Cardiol 55: 1560, 1985
Subaortic stenosis, the univentricular heart, and banding of the pulmonary artery: an analysis of the courses of 43 patients with univentricular heart palliated by pulmonary artery banding.

R M Freedom, L N Benson, J F Smallhorn, W G Williams, G A Trusler and R D Rowe

_Circulation_. 1986;73:758-764
doi: 10.1161/01.CIR.73.4.758

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1986 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/73/4/758