SUMMARY Four patients with tetralogy of Fallot, three of whom had congenital pulmonary atresia, were treated by initial palliative operations to enlarge left and right pulmonary arteries which were considered too small for complete repair. Two to four years later the right and left pulmonary arteries had enlarged sufficiently to allow complete repair.

DIFFUSELY VERY SMALL right and left pulmonary arteries, which occur infrequently, are not amenable to direct surgical enlargement as is the main pulmonary artery, and thus are potentially a contraindication to complete repair of classical tetralogy of Fallot and tetralogy of Fallot with pulmonary atresia, as well as other malformations with severe pulmonary stenosis or atresia. For some years, surgeons have proposed that increasing pulmonary blood flow by patch graft enlargement of the pulmonary valve ring (leaving the ventricular septal defect open) or by a systemic-pulmonary artery shunt would increase the size of the right and left pulmonary arteries sufficiently to allow later definitive repair. The truth of this proposal has not been well documented.

We present four patients in whom enlargement of diffusely small arteries has occurred after palliative operations which increased pulmonary blood flow and presumably intraluminal pulmonary artery pressure. Three of the four have now undergone successful complete repair.

Material and Methods

Four patients, ranging in age from 1.7 to 6.1 years, have undergone initial palliative operations to enlarge very small right and left pulmonary arteries. All four patients had tetralogy of Fallot, one with severe stenosis of the right ventricular infundibulum, pulmonary valve and pulmonary valve ring, and a very small main pulmonary artery, and three with congenital pulmonary atresia. One of the patients with pulmonary atresia had nonconfluent right and left pulmonary arteries and previously had undergone a side-to-end ascending aortic-right pulmonary artery anastomosis (table 1). The other patients had confluent right and left pulmonary arteries.

In one patient (case 2) the palliative operation was done using cardiopulmonary bypass, profound hypothermia to 26°C, and low flow (0.5 L/min/m²) during the repair. An incision in the right ventricular infundibulum was carried across the pulmonary valve ring and to the bifurcation of the main pulmonary artery. The parietal and portions of the septal band were excised, the free wall of the right ventricle mobilized, and most of the pulmonary valve excised. A precollated, measured woven dacron patch was sewed into the incision to enlarge the entire area. In case 1 ascending aorta to both right and left pulmonary artery anastomoses were done through a median sternotomy, after finding the left and right pulmonary arteries too small for the planned repair. Cases 3 and 4 had end-to-side subclavian-pulmonary artery (Blalock-Taussig) anastomoses.

All patients were studied by cardiac catheterization and selective angiocardiography before the palliative operation, and two to five years after it (table 2).

Results

The right and left pulmonary arteries were considerably larger at the time of the postoperative study in all four patients (figs. 1-4). We considered them to be large enough to permit complete repair in all four.

Three of the four patients have successfully undergone complete repair including repair of the ventricular septal defect (tables 1 and 2). In case 1, a heterograft-valved external conduit between right ventricle and the confluence of the pulmonary arteries was used in the repair. Peak pressures after repair were 50 mm Hg in pulmonary artery, 75 in right ventricle, and 85 in left ventricle. In case 2, at the final repair the palliative outflow patch was replaced with a new patch.
extending across the stenotic origin of the left pulmonary artery. Peak pressures after repair were 65 mm Hg in right ventricle and under the patch-graft, 95 in the left ventricle. In both patients, very large aorto-pulmonary collateral (or bronchial) arteries were ligated at the time of complete repair. In case 3, the large collaterals were not disturbed at the time of repair.

Discussion

Left and right pulmonary arteries so small as to preclude primary complete repair of the tetralogy of Fallot (and other malformations) occur infrequently. Between January 1967 and January 1977 these four cases (plus three additional patients in whom palliative patch-graft enlargement across the pulmonary valve ring has successfully been done recently without restudy) were encountered among 515 patients with tetralogy of Fallot with or without pulmonary atresia undergoing surgical treatment. (No such patients were denied operation during this period except a few in whom no pulmonary arteries could be identified in either right or left lung by angiocardiographic study done with special techniques.) Two of these seven patients had classical tetralogy of Fallot with very small right and left pulmonary arteries. This represents 0.5% of the 429 patients with classical tetralogy of Fallot undergoing complete repair during this period. Five of the patients had tetralogy of Fallot and congenital pulmonary atresia, representing 13.2% of the 41 patients with this malformation undergoing complete repair. This reflects the tendency for the degree of hypoplasia of the peripheral pulmonary arteries to be positively correlated with the degree of underdevelopment of the right ventricular outflow tract and pulmonary valve ring.

Norwood has reported a case with severely hypoplastic pulmonary arteries in an infant, but our patients and most of those reported by others have been older than this. The hypoplasia may therefore be congenital in some patients, but in others may be in part the result of a chronically low distending pressure secondary to the low pulmonary blood flow. Patients with the tetralogy of Fallot and small

Table 1. Tetralogy of Fallot with Small Pulmonary Arteries

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Anatomy of pulmonary stenosis or atresia</th>
<th>Aorto-pulmonary collateral flow</th>
<th>Definitive repair with closure of VSD</th>
<th>Repair details</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>pulmonary atresia, absent main PA, confluent R &amp; L PAs</td>
<td>large, discrete &quot;bronchial a.,&quot; upper descending aorta</td>
<td>closure &quot;bronchial a.,&quot;</td>
<td>75/85</td>
</tr>
<tr>
<td>2</td>
<td>severe stenosis infundibulum and PV ring, small main PA, confluent R &amp; L PAs</td>
<td>large, discrete rt. paramediastinal collateral artery</td>
<td>RV—confluence of PAs</td>
<td>50/85</td>
</tr>
<tr>
<td>3</td>
<td>pulmonary atresia, absent main PA, confluent R &amp; L PAs</td>
<td>large, discrete &quot;bronchial a.,&quot; upper descending aorta</td>
<td>closure large collateral artery, patch-graft enlargement across PV ring and across stenosis, origin LPA</td>
<td>65/95</td>
</tr>
<tr>
<td>4</td>
<td>pulmonary atresia, absent RV infundibulum and main PA, nonconfluent RPA and LPA, previous side-to-end ascending aorta—RPA anastomosis</td>
<td>diffuse, small</td>
<td>Patch-graft enlargement across PV ring</td>
<td>50/100</td>
</tr>
</tbody>
</table>

Abbreviations: PA = pulmonary artery; RPA, LPA = right, left pulmonary artery; RV = right ventricle; PV = pulmonary valve; PA/LV = systolic pressure right ventricle/systolic pressure left ventricle; VSD = ventricular septal defect.

Table 2. Tetralogy of Fallot with Small Pulmonary Arteries

<table>
<thead>
<tr>
<th>Case no. and date of birth</th>
<th>Pre-palliation catheterization</th>
<th>Palliative operation</th>
<th>Recatheterization</th>
<th>Repair</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 3/22/74 10/2/70</td>
<td>3/27/74 3.5 RPA &amp; LPA = 3 mm dia</td>
<td>3/26/74 3.5 systemic PA anastomosis, LPA &amp; RPA 4 mm dia</td>
<td>3/29/76 5.5 marked but uneven enlargement RPA &amp; LPA RPA 5 mm diam, stenosis origin LPA</td>
<td>1/28/76 4.0</td>
</tr>
<tr>
<td>2 8/13/73 2/11/72</td>
<td>10/17/73 1.7 RPA &amp; LPA = 2 mm diam</td>
<td>9/30/75 3.6 patch-graft enlargement across PV ring</td>
<td>1/28/76 4.0</td>
<td></td>
</tr>
<tr>
<td>3 11/5/68 4/22/68</td>
<td>9/8/72 4.4 systemic-PA anastomosis, LPA 3 mm diam</td>
<td>8/19/76 8.3 LPA &amp; prox. RPA 5-6 mm diam, localized narrowing, both branches RPA</td>
<td>6/10/77 9.2</td>
<td></td>
</tr>
<tr>
<td>4 7/26/72 7/6/66</td>
<td>7/31/72 6.1 LPA = 2 mm diam</td>
<td>7/12/76 10.0 LPA 5-6 mm diam</td>
<td>Scheduled</td>
<td></td>
</tr>
</tbody>
</table>
peripheral pulmonary arteries should be considered promptly for a surgical procedure to increase pulmonary blood flow and hopefully to maintain or increase the size of the pulmonary arteries.

The identification of patients whose pulmonary arteries are too small to permit complete repair is an imprecise matter at the moment. “Too small” means that the resistance to flow through them would be so great as to
result in inordinately high pressure in the right ventricle and main pulmonary artery after repair and in low cardiac output. The low pulmonary blood flow and the proximal stenotic lesions in the right ventricle and pulmonary valve make direct measurement of total pulmonary resistance technically difficult. Angiocardiography identifies the small size of the right and left pulmonary arteries, but does not define the limits of “too small.” McGoon and colleagues consider, apparently empirically, that in order for complete repair to be considered, “the combined cross-sectional area of the right and left pulmonary arteries at the hilus must be greater than one half the cross-sectional area of the descending thoracic aorta.” This and other angiographic methods have the disadvantage that the size of the right and left pulmonary arteries post-repair with a normal distending pressure may be larger than their apparent size in vivo preoperatively. Thus, the data of McGoon et al. indicate that the size of the right and left pulmonary arteries as determined by angiocardiography can double within two weeks of an operation resulting in increased pulmonary blood flow. This increase must be solely the result of the increased flow and distending pressure.

Jarmakani and colleagues have presented data on the normal size of the right pulmonary artery, determined angiocardiographically, related to body surface area. These authors point out the difficulty in relating angiographic measurements to measurements at autopsy and thus by implication at operation, probably again because of the matter of distending pressure. Normal values for the size of the pulmonary valve ring are available, and we have determined the functional significance of its size measured at operation in patients with tetralogy of Fallot. Such information is not available for the right and left pulmonary arteries.

In practice, extremely small size of well-visualized right and left pulmonary arteries (such as shown in fig. 2A) makes it probable that these vessels are “too small” to permit complete repair. When the arteries are diminutive but the probability that they are “too small” is not as great as this, we believe it best to perform complete repair including an adequate outflow patch across the pulmonary valve ring. We recommend that the surgeon measure the right ventricular/left ventricular systolic pressure ratio after bypass but before decannulation, and if the pressure ratio is greater than 1.0 re-establish cardiopulmonary bypass and make a large hole in the center of the patch used to close the VSD. (A right ventricular/left ventricular systolic pressure ratio greater than 0.65 is undesirable, but in the instance described accepting one up to 1.0 is a better alternative than any other.)

Whether such a palliative patch-graft enlargement across the pulmonary valve ring is preferable to a systemic-pulmonary artery shunt for the purpose of enlarging the right and left pulmonary arteries is controversial. We agree with Gill et al. that it probably is, because when this is done properly flow goes freely into both right and left pulmonary arteries. Of course, palliative anastomotic operations are also reasonable options, as this experience shows. However, with side-to-side ascending aorta-right pulmonary artery anastomosis, a very small right pulmonary artery may kink and reduce or abolish shunt flow through the left pulmonary
When the Blalock-Taussig shunt is used, very small right or left pulmonary arteries may be elevated sufficiently by the anastomosis also to kink the artery, particularly in infants. In patients with pulmonary atresia and nonconfluent right and left pulmonary arteries, bilateral systemic-pulmonary artery shunts of the Blalock-Taussig type (using Laks and Castaneda's modification on the side of the aortic arch) are probably the best option.

Norwood has reported an instance in which he used a palliative right ventricular outflow reconstruction in a patient with tetralogy of Fallot and very small right and left pulmonary arteries, but he also repaired the ventricular septal defect with a perforated patch. The advantage is that the perforation may close spontaneously, and if it does not the second operation is simpler than otherwise. We have not done this routinely in order to keep the initial palliative
WEAR IN BEALL 104 PROSTHESIS/Silver, Wilson

operation as simple as possible, but in two recent cases we have perforated the patch secondarily in the circumstances described earlier.

We have not closed the large aorto-pulmonary collateral vessels at the initial palliative procedure and in case 3 did not close them at the final repair. This remains a controversial matter.

We do not know the rate at which the pulmonary arteries enlarge after these procedures. The magnitude of the increase in size late postoperatively indicates that true growth has occurred, in addition to the initial enlargement from the immediately increased intraluminal pressure. The persistence of the narrowing in the first part of the left pulmonary artery in case 2, and in the two divisions of the right pulmonary artery in case 3, indicates that not all parts of all pulmonary arteries necessarily enlarge as a result of this stimulus. Gerbode, in reporting three patients in whom palliative patch-graft enlargement of the pulmonary-valve ring has successfully enlarged the right and left pulmonary arteries and allowed later complete repair,7 has described an additional patient in whom enlargement did not result.

References

2. McGoon DC, Baird DK, Davis GD: Surgical management of large bronchial collateral arteries with pulmonary stenosis or atresia. Circulation 52: 109, 1975

The Pathology of Wear in the Beall Model 104 Heart Valve Prosthesis

MALCOLM D. SILVER, M.D., AND GREGORY J. WILSON, M.D.

SUMMARY We examined 13 Beall model 104 prostheses recovered at surgery or autopsy 14 to 84½ months after insertion and observed the pattern of prosthesis wear. We defined wear as “mild” when the disc was notched but neither the metal of the struts was exposed nor the cloth seat torn. In “moderate” wear the disc was notched, the Teflon coating of the struts was worn away exposing the underlying metal but the cloth of the seat was not torn. The cloth seat was torn in “severe” wear, exposing the metal seat and causing a different pattern of disc erosion. In one case this allowed the disc to tilt into the valve lumen and, later, to escape from its cage. Most of the prostheses studied showed “moderate” or “severe” wear. We believe that some degree of wear is inevitable. Clinical signs and symptoms were not specific but a rough correlation existed between the severity of hemolysis, as indicated by serum LDH levels, and the degree of prosthesis wear. When wear was “severe,” the results of cardiac catheterization studies usually mirrored the change, but there were a few exceptions. All patients with the Beall model 104 prosthesis may eventually develop “severe” wear, and we recommend regular reassessment with a view toward prosthesis replacement.

ALTHOUGH AN ARTIFICIAL HEART VALVE may be well designed, constructed of materials thought appropriate and be tested in the laboratory, it may behave in an unexpected fashion years after its insertion in man. When this occurs, the manufacturer either withdraws the prosthesis from use or modifies it to overcome the problem. Teflon,* used both to form the disc and to coat the cage in the model 104 Beall disc valve prosthesis, wears with time and has now been replaced by pyrolytic carbon. Many patients had this model prosthesis inserted; approximately 3000 were sold between October 1969 and September 1975 (personal communication from Surgitool Division of Travenol Laboratories). Individuals with this older prosthesis thus form a special group that must be observed closely to avoid untoward events. In this paper, we present the pathology of wear in this prosthesis and discuss clinical parameters that may help a physician decide when to replace it.

Materials and Methods

At the Toronto General Hospital, between 1969 and 1973, Beall model 104 heart valve prostheses were inserted in the mitral area in 90 patients. Between 1973 and March 1977, 13 prostheses showing varying degrees of wear were studied in our laboratory. Each was examined in detail, its degree of wear established and it was photographed. Histological sections prepared from the organs of patients at autopsy were stained with hematoxylin and eosin and examined for Teflon

*DuPont tradename for a tetrafluorethylene resin

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Received April 26, 1977; revision accepted May 23, 1977.
The enlargement of small pulmonary arteries by preliminary palliative operations.
J W Kirklin, L M Barger, Jr and A D Pacifico

Circulation. 1977;56:612-617
doi: 10.1161/01.CIR.56.4.612
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

The online version of this article, along with updated information and services, is located on
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