Pulmonary Atresia or Severe Stenosis and Coronary Artery-to-Pulmonary Artery Fistula

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SUMMARY

Five patients have had the common features of pulmonary atresia or severe stenosis associated with a septal defect and a coronary artery-to-pulmonary artery (CA-PA) fistula. Four had pulmonary valvular atresia, and one had severe pulmonary stenosis. In all five, the CA-PA fistula contributed the principal component of the pulmonary blood flow. Cyanosis, continuous murmur, right ventricular hypertrophy, and decreased or normal pulmonary vascularity were frequent clinical manifestations, but angiocardiography was required to establish the diagnosis. The fistula was a side-to-side communication between the left coronary artery and the main pulmonary artery in each of the five patients. The dilated proximal coronary artery connecting the aortic root with the pulmonary artery gave a unique angiocardiographic appearance distinguishing it from truncus arteriosus or aorticopulmonary window. It is suggested that this unique angiocardiographic appearance be called an “aorticopulmonary tunnel.” The surgical repair of this anomaly is closely related to that for severe tetralogy of Fallot and should involve a comparable risk and achieve a comparable result.

Additional Indexing Words:
Pulmonary atresia  Coronary AV fistula  Pulmonary stenosis

A THE MAYO Clinic, there have been five patients who had pulmonary valvular atresia (four patients) or severe pulmonary stenosis (one patient) in whom the pulmonary blood flow was principally provided by a fistula between a coronary artery and the main pulmonary artery just beyond the atretic or stenotic segment. One of these patients has been previously described.1 We are aware of four similar cases previously reported: two patients with pulmonary atresia2,3 and two patients with pulmonary stenosis.4,5 This report summarizes the unique aspects of this unusual anomaly as encountered in the five patients seen at the Mayo Clinic.

Observations

Clinical Observations (Table 1)

The patients' ages ranged from 2½ years to 21 years. Four were females and one was a male. Only one was asymptomatic and acyanotic at examination; two were minimally cyanotic, and two were moderately-to-severely cyanotic. The second heart sound was single in all patients with pulmonary atresia (cases 1–4), and it was widely split in the only patient with pulmonary stenosis (case 5).
A loud continuous murmur was heard in each patient, with the location of maximal intensity varying considerably. The murmur was associated with a thrill in three of the five patients.

Electrocardiographic Findings (Table 2)

The electrocardiogram in all five patients showed normal sinus rhythm and normal atrioventricular conduction time. Right-axis deviation and isolated right ventricular hypertrophy were present in four of the five patients. The electrocardiogram of one patient (case 4) was exceptional in that it showed combined ventricular hypertrophy with marked dominance of the left ventricle. The patient had the highest calculated pulmonary blood flow of 8.9 liters/min/m² and aortic insufficiency which contributed to the left ventricular hypertrophy seen on his electrocardiogram.

Roentgenographic Findings (Table 2)

The cardiothoracic ratio on the chest roentgenograms ranged from 0.50 to 0.58. The aortic arch was on the left in three, and on the right in two patients. The pulmonary vascular markings were normal in three patients, decreased in one, and increased in one. One patient (case 2) had a bronchial collateral pattern of pulmonary circulation on the plain chest roentgenogram.

Cardiac Catheterization

The right ventricular pressure was at systemic levels in all the patients. In none of the patients with pulmonary atresia was the pulmonary artery entered by the catheter. The pulmonary artery was entered via the right ventricle in one patient with pulmonary stenosis (case 5).

Desaturation of systemic arterial blood was present in each of the five patients. The systemic arterial oxygen saturation ranged from 91% in case 5 to 58% in case 3. The highest systemic saturation (91%) was present in the patient with pulmonary stenosis (case 5). The highest systemic saturation in the patients with pulmonary atresia was 89% in case 4.

Angiographic Studies and Anatomic Diagnosis

The correct diagnosis was established by selective angiography into the right ventricle in two patients and into the right ventricle and the left coronary ostium in one patient. In two patients the correct diagnosis was made during surgery.

Four patients had pulmonary valvular atresia (cases 1–4) and one had pulmonary stenosis (case 5). All had ventricular septal defects. Four of the five patients had angiographic evidence of severe hypoplasia of the main pulmonary artery, and one

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Table 1

Clinical Manifestations in Five Cases of Coronary Artery-to-Pulmonary Artery Fistula Associated with Pulmonary Atresia or Severe Stenosis

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex &amp; age (yr)</th>
<th>Exercise tolerance</th>
<th>Cyanosis</th>
<th>Clubbing</th>
<th>Second heart sound</th>
<th>Murmur</th>
<th>Thrill</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F, 43/4</td>
<td>Decreased</td>
<td>Minimal</td>
<td>No</td>
<td>Single</td>
<td>Continuous</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>F, 83/2</td>
<td>Decreased</td>
<td>Yes</td>
<td>Yes</td>
<td>Single</td>
<td>Continuous</td>
<td>Yes</td>
</tr>
<tr>
<td>3†</td>
<td>F, 23/2</td>
<td>Poor</td>
<td>Moderate</td>
<td>Yes</td>
<td>Single</td>
<td>Continuous</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>M, 14</td>
<td>Decreased</td>
<td>Mild</td>
<td>Minimal</td>
<td>Single</td>
<td>Continuous</td>
<td>Yes</td>
</tr>
<tr>
<td>5</td>
<td>F, 21</td>
<td>Asymptomatic</td>
<td>No</td>
<td>No</td>
<td>Split</td>
<td>Continuous</td>
<td>Yes</td>
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</tbody>
</table>

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Table 2

Electrocardiographic and Roentgenographic Findings in Five Cases of Coronary Artery-to-Pulmonary Artery Fistula Associated with Pulmonary Atresia or Severe Stenosis

<table>
<thead>
<tr>
<th>Case</th>
<th>Mean frontal axis rotation°</th>
<th>Hypertrophy</th>
<th>Cardiacoriches ratio</th>
<th>Pulmonary vasculature</th>
<th>Aortic arch</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>+110</td>
<td>RVH</td>
<td>0.51</td>
<td>Normal</td>
<td>R</td>
</tr>
<tr>
<td>2</td>
<td>+150</td>
<td>RVH</td>
<td>0.53</td>
<td>Normal</td>
<td>L</td>
</tr>
<tr>
<td>3†</td>
<td>+130</td>
<td>RVH</td>
<td>0.56</td>
<td>Decreased</td>
<td>L</td>
</tr>
<tr>
<td>4</td>
<td>+60</td>
<td>CVH</td>
<td>0.58</td>
<td>Increased</td>
<td>R</td>
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<tr>
<td>5</td>
<td>+120</td>
<td>RVH</td>
<td>0.50</td>
<td>Normal</td>
<td>L</td>
</tr>
</tbody>
</table>

Abbreviations: RVH = right ventricular hypertrophy; CVH = combined ventricular hypertrophy; R = right; L = left.

*Clockwise rotation.
Case 2. Angiogram showing injection into right ventricle. Anteroposterior view. Note aorticopulmonary tunnel-dilated proximal part of coronary artery (arrow) and preferential filling of ascending aorta via ventricular septal defect.

Case 2. Simultaneous lateral view of previous injection (fig. 1), with filling of proximal dilated coronary artery (arrow).
Figure 3

Case 2. Subsequent frame of biplane angiogram (figs. 1, 2), showing filling of both aorta and pulmonary artery. Note hypoplastic pulmonary artery and anomalous blood vessel from left subclavian artery supplying left upper lobe. Nature of ascending aorta-pulmonary artery communication is not apparent without documentation of the aortopulmonary tunnel as seen in figures 1 and 2. Patent ductus arteriosus is excluded.

(case 4) had a normally sized main pulmonary artery on the angiogram.

In all five the communication was side-to-side between the left coronary artery and the main pulmonary artery. In each the proximal part of the coronary artery was dilated to the same size as the hypoplastic pulmonary artery.

Selective angiography in all patients showed a distinct pattern. This pattern was an aortopulmonary tunnel consisting of the proximal dilated coronary artery which formed a communication between the ascending aorta and the pulmonary artery (figs. 1–3). This angiographic feature is pathognomonic and excludes the diagnosis of truncus arteriosus or aortopulmonary window.

Surgical Treatment

Total correction was carried out in all five patients. One patient (case 3) required a palliative Blalock-Taussig anastomosis at 2½ years of age, prior to total correction which was carried out 3 years later.

In all, the coronary artery-pulmonary artery fistula was ligated, oversewn, or disconnected, and the ventricular septal defect was closed with a patch. In two of the five patients, the right ventricular outflow tract was reconstructed using either a pericardial tube graft (case 3) or an aortic homograft including the aortic valve (case 1). In three (cases 2, 4, and 5), a longitudinal ventriculotomy was extended across the atretic pulmonary valve and a pericardial gusset was inserted. One patient (case 1) died following surgery. Three patients (cases 2, 3, and 4) have evidence of residual pulmonary insufficiency.

One patient (case 3) underwent cardiac catheterization 4 years after operation because of decreasing exercise tolerance and an increase in cardiac size. The cardiac catheterization showed systemic pressure in the right ventricle and the pulmonary artery due to peripheral pulmonary arterial stenosis.

Discussion

The incidence of coronary artery fistulas communicating with the various chambers of the heart is about 0.2–0.4% of congenital heart disease. By supplementing the extensive review of congenital anomalies of the coronary arteries by Ogden, we collected data on 310 cases of coronary artery fistula. Of these, 276 were without additional major valvular...
malformations (primary fistula),\textsuperscript{8} and 34 were associated with major valvular malformations (secondary fistula) (table 3). Secondary coronary artery fistula constitutes 11\% of the total number reported in the literature. Most reported instances of secondary coronary artery fistula (73\%) involve a communication between the coronary artery and the right ventricle in the presence of pulmonary valvular atresia or severe pulmonary valvular stenosis. Such hearts may or may not have a ventricular septal defect.

**Embryologic Considerations**

Grant\textsuperscript{40, 41} and Edwards\textsuperscript{8} suggested that coronary artery fistula in the presence of severe valvular malformations is secondary to the persistence of the primitive intramyocardial sinusoids. The data in table 3 support this developmental analysis, inasmuch as the incidence of communication between the coronary artery and the right ventricle is greatly increased in the presence of pulmonary atresia in comparison to that in primary coronary artery-pulmonary artery fistulas. A similar trend is noted in patients with coronary artery-left ventricle communication in the presence of aortic atresia, although the total number involved is small. If such an association between pulmonary atresia and the development of coronary artery-pulmonary artery fistula exists, it is less evident insofar as recorded numbers of cases in comparison with coronary artery-right ventricle communications.

Recently Ogden\textsuperscript{42} offered a theory for the development of the coronary arteries that suggests a dual origin from both the proximal aortic buds and the distal retiform vascular network. Such a theory may explain the various anatomic patterns encountered in coronary artery-pulmonary artery fistulas with or without pulmonary atresia. In 45 patients with primary coronary artery-pulmonary artery fistulas previously reported,\textsuperscript{4} the communication in 15 was via a retiform vascular network consisting of multiple small vessels connecting the aorta to the pulmonary artery. None of the five patients reported by us or the four patients previously reported\textsuperscript{2-5} with coronary artery-pulmonary artery fistula and pulmonary valvular atresia or stenosis had a retiform vascular communication. This significant difference ($P = 0.04$) suggests that, in patients with coronary artery-pulmonary artery fistula and associated pulmonary atresia, a preferential development of one branch of the embryologic retiform network occurs.

**Clinical Considerations**

In two patients, the continuous murmur may have resulted from causes other than the coronary artery-pulmonary artery fistula. One (case 2) had an anomalous systemic vessel to the left upper lobe, and one patient (case 4) had dilated bronchial arteries.

Some authors\textsuperscript{12, 43, 44} emphasized the presence of a coronary artery “steal syndrome” in patients with coronary artery-to-pulmonary artery communications, such as coronary

### Table 3

<table>
<thead>
<tr>
<th>Chamber of termination</th>
<th>Primary*</th>
<th>Secondary</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
<td>Pulm atresia</td>
<td>Pulm stenosis</td>
<td>Aortic atresia</td>
</tr>
<tr>
<td>Right atrium</td>
<td>79</td>
<td>29</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<tr>
<td>Right ventricle</td>
<td>106</td>
<td>38</td>
<td>21</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>58</td>
<td>21</td>
<td>3</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>9</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Left atrium</td>
<td>14</td>
<td>5</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Other</td>
<td>10</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>276</td>
<td>100</td>
<td>25</td>
<td>6</td>
<td>3</td>
</tr>
</tbody>
</table>

\*Without major congenital malformations.
artery-to-pulmonary artery fistula or anomalous origin of the left coronary artery from the pulmonary artery. However, none of the patients in our report experienced angina. This might be explained partially on the basis of the younger age group of the patients with coronary artery-pulmonary artery fistula and associated pulmonary atresia, and partly on the basis of the limited activities of most of these patients.

Some misinterpretations of the findings at the initial catheterization obscure the correct diagnosis in most cases. In none of the patients with pulmonary atresia was the pulmonary artery entered through the coronary artery-pulmonary artery fistula. At times, in patients believed to have truncus arteriosus, type I, or aorticopulmonary window, repeated attempts to advance the catheter to the distal pulmonary artery resulted in the repeated slipping of the catheter from the "main pulmonary artery" (proximal dilated coronary artery) into the distal left coronary artery. This should suggest the possibility of coronary artery-pulmonary artery fistula. The difficulties encountered in passing the catheter into the pulmonary artery arise from the tortuosity of the coronary artery and the side-to-side nature of the fistula. However, it is probably best not to pass the catheter through the fistulous communication, especially in severely cyanotic patients, because the presence of the catheter might further restrict pulmonary blood flow and might be life threatening.

In two of the patients, the systemic pressure in the proximal dilated coronary artery was thought at first to represent the main pulmonary arterial pressure. In one of these, the hemodynamic calculations based on such an assumption fallaciously showed that the patient had severe pulmonary vascular obstructive disease and excluded surgical correction, but the presence of a continuous murmur led to the correct interpretation of the catheter position. In both patients, the true main pulmonary arterial pressure was recorded at surgery and was 22/17 mm Hg and 25/20 mm Hg. At the present time, no patient has been described with pulmonary valvular atresia, coronary artery-pulmonary artery fistula, and pulmonary vascular obstructive disease.

Selective angiography, preferably into the aortic root or the proximal part of the coronary artery,6 with demonstration of an aorticopulmonary tunnel is the single most important test in the diagnosis of pulmonary valvular atresia and coronary artery-pulmonary artery fistula.

Surgical Considerations

The repair of this condition involves three steps. First, the coronary artery-pulmonary artery fistula must be closed. In order to avoid dissection of the coronary arteries, particularly the upstream dilated parent vessel with the possible consequent injury or interruption of important branches, we prefer to oversew the fistula from within the pulmonary artery. It is readily accessible, and its margins can be approximated without tension. Second, the septal defect is corrected in the usual manner.47

Third, the pulmonary atresia or stenosis is relieved. In tetralogy with severe pulmonary stenosis or even localized valvular or infundibular pulmonary atresia, surgeons vary as to their choice of technic for relieving the outflow tract obstruction;48 that is, whether to extend the ventriculotomy through the stenotic or atretic segment and insert a pericardial patch graft, or to separate the pulmonary artery from the heart and interpose a homograft great artery and its contained semilunar valve.49

In other more complex conditions where there is complete discontinuity between the right ventricle and the pulmonary artery,50 as in truncus arteriosus, pulmonary arterial atresia, or transposition with pulmonary stenosis, there is no choice but to insert a conduit such as an aortic homograft between the right ventricle and the pulmonary artery. The presently reported condition lies between these two groups. Because pulmonary resistance is low in most of these patients, there is no more need for a competent pulmonary valve than after repair of the severe tetralogy. We prefer pericardial outflow-tract patch
reconstruction whenever this will suffice to relieve the obstruction, rather than insertion of an aortic homograft.

For patients in whom the coronary artery-pulmonary artery fistula is too small to provide adequate pulmonary blood flow, and who require surgical aid before the age of 4 or 5 years, we prefer a systemic-to-pulmonary anastomosis (such as a Blalock anastomosis) and defer the definitive operation until the age of 5 years, or a little older.

The morbidity and mortality rates and late result for correcting this deformity and the long-term result currently should be similar to those for the repair of severe tetralogy.

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