Pulmonary Valve Atresia with Intact Ventricular Septum

Report of a Case with Long Survival and Pulmonary Blood Supply from an Anomalous Coronary Artery


SUMMARY
A case of pulmonary valve atresia with intact ventricular septum with long survival in the absence of a patent ductus arteriosus, is presented. The patient, alive at 21 years of age, has the anterior descending branch of the left coronary artery originating from the pulmonary artery. Thus, the pulmonary blood supply is obtained from the right coronary artery via intercoronary anastomoses. No similar case has been reported previously.

Additional Indexing Words:
Selective coronary angiography

PULMONARY valve atresia with intact ventricular septum is a rare type of congenital cardiac malformation. Keith et al. assess the incidence at about 1% of the total group of congenital cardiac malformations. The vast majority of patients die in early infancy. Those who survive for a longer period have, almost invariably, a widely patent ductus arteriosus. The patient whose findings are presented here is considered to be unique in that, alive at 21 years of age, the ductus arteriosus is closed and the pulmonary arteries are supplied solely from the right coronary artery through an anomalous anterior or descending branch of the left coronary artery which arises from the pulmonary artery.

Case Report
G.B., a 21-year-old South Indian male, was born normally to nonconsanguineous parents after an uncomplicated pregnancy. The child did not suck well for the first 2 weeks of life but the parents stoutly deny any cyanosis or respiratory distress at this time. Thereafter, the child fed normally but during the first 6 months he cried extremely frequently, so much so that the parents consulted a doctor who diagnosed colic and advised gripe water. There is no record of the cardiac findings at that time, but no cyanosis was noted. After 6 months, the child appeared to develop fairly normally, walking and speaking by the age of 1½ years. Although no dyspnea or cyanosis was present, the child tended to refrain from playing, preferring to sit quietly in the house.

Only at the age of 10 years was the gradual onset of dyspnea on exertion and blueness of the fingers and toes noted. Thereafter, the mother noticed that the child tended to squat down after walking a furlong or so, and at these times his color was darker. He never lost consciousness and there was no chest pain. Within the past year, the patient's symptoms increased and he sought treatment elsewhere; he was referred to Christian

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Medical College Hospital, Vellore, for further investigations and treatment.

Clinical examination revealed a poorly grown individual, of rather low intelligence, weighing 40 kg. There were no external congenital deformities. There was moderate clubbing and central cyanosis. There was no edema. The jugular venous pressure was moderately elevated with an a wave visible to 6 cm above the sternal angle. Pulse was 80 beats per min, regular and of average volume. Blood pressure was 96/60 mm Hg in the right arm. The apical impulse was at the midclavicular line in the fifth left intercostal space and was tapping in character. A moderate parasternal lift was felt. Auscultation revealed a normal first sound, a single, loud second sound, and an ejection sound at the left sternal border. There was a fairly loud (III/VI) continuous murmur best heard at the pulmonary area but heard well at the lower sternal border and also conducted well beyond the precordium. This murmur became louder (IV/VI) on inspiration. No separate systolic or diastolic murmurs could be detected.

The lung fields were clear clinically and the liver was palpable 1.5 cm below the costal margin. The other systems were normal.

Hemoglobin was 21.8 g/100 ml; packed cell volume was 74%.

The electrocardiogram (fig. 1) was interpreted as showing right and left atrial enlargement. The frontal plane QRS axis was +114°. The chest leads revealed very deep S waves in V1 to V5 consistent with left and right ventricular hypertrophy.

Chest roentgenogram (posteroanterior view, fig. 2) showed a cardiothoracic ratio of 50% with rounded left cardiac border, enlarged right atrium, normal aortic and pulmonary artery shadows, and oligemic lung fields.

Cardiac Catheterization

Cardiac catheterization was carried out through the right median antecubital vein and the right brachial artery. Table 1 shows the pressures and oxygen saturation obtained. It was possible to enter the right ventricle (RV) with the venous catheter but the catheter could not be passed into the pulmonary artery (PA) or aorta. The catheter passed readily from the right atrium (RA) through an atrial septal defect to the left atrium (LA) and several pulmonary veins. From the left atrium, the catheter was passed to the left ventricle (LV). The right ventricular pressure was consistently higher than that of the left ventricle and the pressure tracing had the peaked configuration of pulmonary obstruction with intact interventricular septum.

Cineangiography was carried out in both the RV and LV in the posteroanterior and left anterior oblique projections. RV injection (fig. 3) revealed a relatively normalized heavily trabeculated ventricular cavity
Table 1

Results of Cardiac Catheterization

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mm Hg)</th>
<th>Oxygen saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>—</td>
<td>47.0; 49.0</td>
</tr>
<tr>
<td>Right atrium</td>
<td>a = 12; x = 3; v = 6; y = 5; m = 5</td>
<td>49.5; 51.5; 53.0</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>180/0–10</td>
<td>47.0; 47.0; 49.0</td>
</tr>
<tr>
<td>Left atrium</td>
<td>a = 12; x = 4; v = 5; y = 3; m = 4</td>
<td>50.0; 64.0</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>120/0–5</td>
<td>59.0</td>
</tr>
<tr>
<td>Brachial artery</td>
<td>115/70; m = 82</td>
<td>—</td>
</tr>
</tbody>
</table>

Abbreviations: a, x, v, and y denote respective points on the atrial pressure trace; m = mean pressure.

with no exit of contrast to the PA and no ventricular septal defect. Contrast regurgitated from the RV to RA and then passed to LA and LV. A number of dilated channels leading from the RV were considered to be dilated sinusoids. LV injection (fig. 4) showed an average-sized LV cavity, normal aortic root, no patent ductus arteriosus (PDA), and a very dilated and tortuous right coronary artery. A small artery was seen at the usual site of the left coronary artery. The later frames of the angiogram showed the right coronary artery dividing into a number of anastomotic channels and joining one large vessel which emptied into a normal-sized pulmonary artery. Selective injection of contrast into the right coronary artery (fig. 5 and 6) demonstrated this anomalous communication more clearly. The appearances were typical of anomalous origin of the left coronary artery from the pulmonary artery of the so-called adult type.3

After several injections of contrast into the right coronary artery, the patient developed fairly severe breathlessness and bronchospasm. The procedure was abandoned, therefore, without attempting to catheterize the artery seen at the usual site of the left coronary artery. The patient completely recovered in the ward after 1 hr.

Discussion

Although the entity of pulmonary valve atresia with intact ventricular septum was described in 1784 by Hunter,4 it is only relatively recently that much interest has been shown in this condition5–11 with the description by Greenwold et al.6 of two anatomical types, one of which, type II, may be treated by pulmonary valvotomy.

The vast majority of children with this condition die within the first month of life.1 However, rarely, cases have survived as long as 21 years without surgery.12 It is obvious

![Figure 3](http://circ.ahajournals.org/)

**Figure 3**

Selective right ventricular (RV) injection of contrast demonstrates fairly normal-sized RV with heavy trabeculation and infundibular “cul-de-sac.” (This and subsequent illustrations are reproduced from single frames of a cineangiogram.)
Selective left ventricular (LV) injection in left anterior oblique projection. There is no patent ductus arteriosus and the greatly dilated right coronary artery is seen. An artery appears at the site of the circumflex branch of the left coronary artery (arrow).

Figure 5

Early (left) and late (right) frames from a selective right coronary cinearteriogram (right anterior oblique projection). Contrast passes from the right coronary artery to the anterior descending branch of the left coronary artery which joins the pulmonary artery, well filled in the later frame.

that, for life to be prolonged for any length of time, blood must reach the pulmonary arteries by an alternative route. The venous blood passes from the RA to the LA through an atrial septal defect or a widely patent foramen ovale and reaches the left side of the heart and the aorta. From there, it must reach the pulmonary artery. In all previously reported cases, with one exception,\textsuperscript{12} this has been achieved by persistence of the ductus arteriosus. There is no theoretical reason, however, why other systemic-to-pulmonary communications cannot fulfill this role. Indeed, it is rather surprising that no case has been found with, for instance, bronchopulmonary anastomoses as the only source of pulmonary blood supply (as is found in cases of “extreme” tetralogy of Fallot or truncus arteriosus type IV). Robicsek and colleagues\textsuperscript{12} reported a case in which the communication was an aortopulmonary window. In the case presented here, the fortuitous presence of anomalous origin of a branch of the left coronary artery from the
pulmonary artery has provided the necessary systemic-to-pulmonary communication and so has allowed the patient to survive to 21 years of age.

It is difficult to assess the accuracy of the history claiming absence of cyanosis until 10 years of age. However, it seems fairly clear that his symptoms did appear to increase at about that age. One may speculate, therefore, on whether the ductus arteriosus had remained patent until that age but then closed down or became thrombosed, as is known to occur,1 leaving the anomalous coronary artery as the sole source of blood for the pulmonary circulation. The history of extreme fretfulness as an infant must also be treated with caution but a proportion of cases with anomalous origin of the left coronary artery from the pulmonary artery give this history.

The majority of patients with an "adult-type" anomalous left coronary artery syndrome have, on clinical examination, a continuous murmur at the left sternal border and at the pulmonary area. In retrospect, the murmur in this patient was completely typical of the condition, although, taking the patient's total presentation, this anomaly was not suspected prior to catheterization.

The chest roentgenographic appearance was somewhat surprising in that the transverse diameter of the heart was not distinctly increased. As the patient is so much older than the usual case of pulmonary valve atresia with intact ventricular septum, it is probably unwise to make strict comparisons, but it is generally considered that the heart shows progressive increase in its transverse diameter in these cases.8, 13 In the patient who survived to 21 years, as previously mentioned,12 the chest roentgenogram showed cardiomegaly of moderate degree. In addition, cases with anomalous origin of the left coronary artery as the sole lesion tend to have cardiac enlargement.

It is generally agreed that angiography is necessary to fully establish the diagnosis in cases of pulmonary valve atresia with intact ventricular septum, and selective cineangiocardiology clearly delineated the abnormalities present in this instance. The unusual feature of absence of a patent ductus arteriosus was well documented on LV injection. On occasion, in our experience and that of others,13 angiography may not clearly outline a ductus when present, but the complete absence of filling of the pulmonary artery by contrast until after the aortic arch was empty of contrast is fairly conclusive proof of the absence of a PDA.

It would appear that the coronary artery anomaly in this particular case does not involve the whole of the left coronary artery. The LV injection of contrast revealed a coronary artery arising from the aortic root at the site of the normal left coronary artery and running in the position of the circumflex branch of that artery. The trunk joining the pulmonary artery ran in the position usually occupied by the anterior descending branch of the left coronary artery. It appears, therefore, that it is this branch of the left coronary artery that is anomalous. Selective contrast injection into the artery arising from the aortic root in the position of the left coronary artery or direct visualization of the coronary arteries at surgery would have been useful to confirm the coronary anatomy, but the former was precluded by deterioration of the patient's condition during cardiac catheterization and he has not yet agreed to surgery.

Anomalous origin of the anterior descending branch of the left coronary artery from the pulmonary artery must, itself, be rare. Effler et al.14 classifying coronary artery anomalies, included such an anomaly (their group A, type 3b) but did not include an example of such a case. Likewise, Wesselhoeft et al.15 in a large review and Ogden16 in a large series did not mention a case.

As noted, the patient has not as yet agreed to surgery. As the case most closely fits into type II of Greenwald's classification, pulmonary valvotomy would appear to be indicated and a total correction of the anomalies present may be possible.

References


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