Aortic-Pulmonary Window

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SUMMARY

Two cases of aortic-pulmonary artery window are described. This is an uncommon condition, which resembles, and should be distinguished from, a large patent ductus arteriosus. Life expectancy of patients with the condition is reduced. Total surgical correction can be performed, preferably with the aid of cardiac-pulmonary bypass.

An aortic-pulmonary window is an isolated window-like communication between the adjacent portions of the ascending aorta and the pulmonary arterial trunk. It is an uncommon, congenital, cardiovascular defect, which is of unusual importance, because the signs and symptoms of the condition resemble those of a patent ductus arteriosus, whereas the surgical approach differs significantly from that used to correct a ductus arteriosus.

The purpose of this article is to describe two cases of aortic-pulmonary window, and to summarize the clinical features and treatment.

CASE PRESENTATIONS

Case 1

This patient was first examined at the Cardiac Clinic of H.F. Verwoerd Hospital at the age of 4 years. She had suffered from repeated respiratory infections since infancy. As a baby she had had difficulty in taking feeds. At the age of 1 year severe emphysema and bronchitis had developed. Her main complaints at the age of 4 years were excessive tiredness and dyspnoea.

There were no signs of congestive cardiac failure or cyanosis on examination. The blood pressure in the arm was 80/50 mmHg. All pulses were present and strong. A heaving cardiac impulse was palpable in the 6th intercostal space to the left of the midclavicular line. A grade 2 over 6 systolic murmur and a long decrescendo diastolic murmer was best heard over the base of the heart to the left of the sternum. The second sound was physiologically split. The pulmonary component was accentuated.

The electrocardiogram showed left atrial enlargement and biventricular hypertrophy.

X-ray pictures of the heart showed a large aorta, prominent pulmonary artery, left ventricular enlargement and increased pulmonary vascular markings.

Cardiac catheterization produced evidence of a left to right shunt at pulmonary artery level and pulmonary hypertension (Table 1). Pulmonary vascular resistance was raised. Selective aortography demonstrated the presence of a patent ductus arteriosus (PDA).

At thoracotomy a small PDA (5 mm in diameter) was found and ligated. The ductus arteriosus was too small to be responsible for the pulmonary hypertension. After the ductus was closed, it was found that the murmurs and clinical findings were largely unchanged. After the operation, a moderate degree of improvement occurred. Respiratory infections became less common.

Summary of Cardiac Catheterization

<table>
<thead>
<tr>
<th>Date</th>
<th>Position of catheter</th>
<th>Pressure (mmHg)</th>
<th>Percentage O₂ saturation</th>
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<tbody>
<tr>
<td>Case 1: 7 January 1964</td>
<td>PA 80</td>
<td>74</td>
<td></td>
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<tr>
<td></td>
<td>RV 80</td>
<td>66</td>
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<td></td>
<td>Aorta 78</td>
<td>100</td>
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<td></td>
<td>LV 78</td>
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</tr>
<tr>
<td>7 July 1970</td>
<td>PA 82</td>
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<td></td>
<td>RV 82</td>
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<tr>
<td></td>
<td>Aorta 85</td>
<td>99</td>
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</tr>
<tr>
<td></td>
<td>LV 83</td>
<td>98</td>
<td></td>
</tr>
</tbody>
</table>

*Date received: 9 August 1972.*
At the age of 10 years she was re-admitted for investigation. She still complained of tiredness on effort, and in addition of precordial pain, not related to effort.

She was not cyanotic. The pulse rate was 80 per minute. The blood pressure in the arms was 110/60 mmHg. All pulses were pounding. On palpation the cardiac impulse was diffuse and hyperdynamic. A grade 4 over 6 harsh pansystolic murmur, with an early systolic ejection click, was heard maximally over the left sternal border, at the level of the 2nd and 3rd intercostal spaces. The second heart sound over the pulmonary area was physiologically split and the pulmonary component was loud. A very soft early diastolic blowing murmur could be heard.

The X-ray findings were essentially the same as those 6 years previously. The electrocardiogram showed biventricular hypertrophy.

Cardiac catheterization was carried out again. The pulmonary artery pressure was still raised, and she still showed evidence of a left to right shunt at pulmonary artery level (Table I). The pulmonary vascular resistance, however, had fallen and the size of the shunt increased.

Selective angiography demonstrated the presence of a communication between the ascending aorta and the pulmonary artery. The pulmonary and aortic valves were present and separate. No other lesions were found.

The diagnosis of an aortic-pulmonary window was made. Surgical correction was successfully carried out with the aid of complete cardiopulmonary bypass. Her pulmonary arterial pressures returned to nearly normal levels postoperatively.

Case 2

This patient had suffered from repeated respiratory infections since the age of 2 months. She became easily tired, perspired excessively, and at times became blue.

On examination at the age of 1 year, she was small for her age, mildly cyanotic and dyspnoeic. Blood pressure was 90/50 mmHg. Pulses were strong. The heart was enlarged with biventricular hypertrophy. A long harsh systolic murmur was best heard over the pulmonary area. The second sound was split with a loud pulmonary component.

The electrocardiogram showed biventricular hypertrophy (Fig. 1). X-ray pictures showed enlargement of all of the chambers of the heart, as well as the aorta and pulmonary artery and increased vascularity of the lungs (Fig. 2).

Cardiac catheterization produced evidence of a bidirectional shunt at pulmonary artery level, increased pulmonary vascular resistance and pulmonary arterial systolic pressure equal to systemic pressure (Table I).

An aortic-pulmonary window was repaired under cardiopulmonary bypass.

**DISCUSSION**

An aortic-pulmonary window is a congenital defect of the aorticopulmonary artery septum. It is commonly single, but two openings may be present. In size it varies from 2 to 50 mm in diameter and is round or oval in shape. The communication is situated a short distance above the coronary arteries.

It is an uncommon condition. In a review of the literature, Lynch et al. in 1969, found only 91 published cases. Cleland stated that at Hammersmith Hospital they had encountered 4 cases among over 200 with ventricular septal defect.

The effect on the circulation is similar to that of a patent ductus arteriosus, resulting in a left to right shunt, increased pulmonary blood flow and an increased volume load on the left heart. Pulmonary hypertension is common, Secondary pulmonary vascular changes may occur as early as the first year of life.

The prognosis without treatment is very poor. Abbott found 10 cases in her series of 1000 autopsies of congenital cardiac defects, whereas the incidence in clinical practice may be more than 1 in 1000 cases of congenital heart disease.

The clinical picture resembles that of a large patent ductus. Infants or children with a low pulmonary
vascular resistance and increased pulmonary flow, will
be small for age, sweat excessively, become easily tired
and short of breath on taking feeds or after exercise,
and suffer from frequent respiratory infections. The
pulses are pounding. Signs of heart failure may be
present. The common murmur is a harsh systolic ejection
type of murmur, best heard at the left sternal border,
more medial and lower than where a patent ductus
arteriosus murmur is commonly heard. A continuous
murmur is not common. Associated with the above,
may be a decrescendo diastolic blowing murmur, due
to pulmonary incompetence, and a systolic ejection click
in patients with pulmonary hypertension and a dilated
pulmonary artery. The second sound over the pulmonary
area is split, and the pulmonary component louder than
normal.

Pulmonary vascular disease will result in diminution
of pulmonary blood flow, and produce cyanosis, right
ventricular hypertrophy and softening of the murmur.

The electrocardiographic changes will depend on the
haemodynamic state, with the spectrum ranging from
left ventricular hypertrophy in cases with a large left
to right shunt, to biventricular and right ventricular
hypertrophy as the pulmonary resistance rises.

Roentgenographic signs will also depend on the degree
of pulmonary vascular resistance. A large heart with an
enlarged left ventricle and left atrium and increased
pulmonary vascularity, will be encountered in cases
with low resistance. In cases where pulmonary vascular
obstruction is present, right ventricular hypertrophy, a
large proximal pulmonary artery and avascular peri-
pheral lung fields, will be found.

The diagnoses can be established only by cardiac
catheterization, where selective angiocardiography is
essential to demonstrate the site of the lesion in the
ascending aorta, thus distinguishing it from a ductus
arteriosus. It is also essential to demonstrate the presence
of separate pulmonary and aortic valves, thus ruling
out a true persistent truncus arteriosus. In addition, it
is important to look for associated defects such as a
PDA, coarctation of aorta, ventricular septal defect,
atrial septal defect, and right aortic arch, which are
not uncommon.7

Surgical correction is best carried out using total
cardiopulmonary bypass.87 Closure of the defect by
external ligation,9 or using clamps,9 has been done, but
it is hazardous and uncertain, unless the lesion is
sufficiently high and does not impinge upon the right
pulmonary artery.9

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