Case reports

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Effect of coexistent coarctation of pulmonary trunk in natural history of complete absence of pulmonary valve with ventricular septal defect

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A case of complete absence of the pulmonary valve in association with a ventricular septal defect and a pulmonary artery coarctation is described. Despite these defects the patient had minimal symptoms, probably because the pulmonary artery coarctation acted as a natural banding and limited the pulmonary regurgitant flow and reduced the left-to-right shunt across the ventricular septal defect. The patient was operated on at the age of 40 and the ventricular septal defect was closed but the pulmonary artery coarctation was left alone.

In view of the long-term survival in this case, it is suggested that pulmonary artery banding would reduce the symptoms and increase the life expectancy of patients with congenital absence of the pulmonary valve and a ventricular septal defect.

Congenital absence of the pulmonary valve was first described by Chevers in 1847 and since then about 60 cases have been reported (Nadas and Fyler, 1972). The large majority of these have an associated ventricular septal defect (Ruttenberg, 1968) especially as an additional feature in Fallot's tetralogy (Miller, Lev, and Paul, 1962). Long-term survival is rare and most patients die in infancy (Gasul, Arcilla, and Lev, 1966). We report a case of complete absence of the pulmonary valve with a ventricular septal defect, with long-term survival and minimal symptoms possibly because of a coexistent coarctation of the pulmonary trunk acting as a natural ‘banding’.

Case report

The patient, a woman, had been known to have a murmur since infancy. She was first seen in this hospital in 1958 at the age of 26 when she presented with a history of gradual onset of dyspnoea and palpitations on effort. At that visit she had no cyanosis or clubbing. There was a giant 'a' wave in the venous pulse, and a moderate lift over the right ventricle could be felt. The pulmonary artery was not palpable. The apex beat was localized to the midclavicular line. At the base, there was a normal first sound, a loud ejection type systolic murmur conducted to the neck, and a loud, high-pitched, half-length early diastolic murmur which was also heard at the axillae and in the interscapular area. The second sound was single. The lung bases were clear and the liver was not palpable. The electrocardiogram showed right ventricular hypertrophy and ventricular ectopic beats. The chest x-ray (Fig. 1) showed cardiac enlargement, a small aorta with a right-sided aortic arch, enormous dilatation of the pulmonary artery branches, and moderate pulmonary plethora. A right heart catheter study at this time showed a large left-to-right shunt at the high ventricular level with a systolic gradient of 30 mmHg (4.0 kPa) on withdrawal from the main pulmonary artery to the right ventricle (Table).

The patient refused surgery, and thereafter was followed in the outpatient department. She remained well with only minimal shortness of breath on exertion. She had an uncomplicated pregnancy in 1960. In February 1974 she was admitted for review. Her only symptoms were some tiredness and easy fatiguability. There was no change in the clinical signs, electrocardiogram, or chest x-ray. The cardiac catheterization data on this admission are shown in the Table. An increase in the gradient from the main pulmonary artery to the right ventricle was seen, and a diagnosis of a high ventricular septal defect with a large left-to-right shunt was reaffirmed.

Surgery was advised and at operation the aorta was found to be small and the right and left pulmonary arteries were enormously dilated and were about two and
Coexistent coarctation of pulmonary trunk in natural history of complete absence of pulmonary valve

FIG. 1 The chest x-ray posteroanterior view.

a half times the size of the ascending aorta. There was a narrowing in the main pulmonary artery about 2 cm above the valve ring. Systolic and diastolic thrills were palpable at this level. The right heart chambers were enlarged, but the left ventricle was quite small. A finger, passed through the right atrial appendage, palpated a normal atrial septum, normal tricuspid valve, and high ventricular septal defect. On total cardiopulmonary bypass and hypothermia the right ventricle was opened transversely in the outflow tract revealing a thickened wall, but no infundibular fibrosis or stenosis. There was complete absence of the pulmonary valve. The diameter of the coarctation was estimated to be about 1.5 cm. The ventricular septal defect was approximately 2 cm in diameter and was situated just below the aortic valve. The defect was closed with multiple interrupted sutures on 'teflon' pledges. No infundibular muscle was resected and the coarctation in the main pulmonary artery was left alone. The patient was discharged after an uneventful recovery two weeks later and has since remained free of symptoms on regular follow-up.

A postoperative right heart catheterization was done in July 1974. The haemodynamic data are given in the Table and a right ventricular angiogram done at this study is shown in Fig. 2.

**Discussion**

Congenital pulmonary regurgitation can be caused by hypoplasia of a cusp (Stintzing, 1889; Ford et al., 1956), bicuspid valve or a quadricuspid valve (Ehrenhaft, 1955; Smith, DuShane, and Edwards, 1959; Ito, Engle, and Holswade, 1961). The anomaly is rarely reported as an isolated lesion (Stintzing, 1889; Kissin, 1936; Ehrenhaft, 1955; Ford et al., 1956; Dickens, Raber, and Goldberg, 1958; Smith et al., 1959) and is usually associated with a ventricular septal defect (Lavenne et al., 1954; Onesti and Harned, 1958; Campeau, Gilbert, and Acierchide, 1961; Vlad, Weidman, and Lambert, 1960; Venables, 1962; Ruttenberg, 1968; Macartney and Miller, 1970), or a Fallot’s situation (Miller et al., 1962). Rarely other complex lesions, such as double outlet right ventricle and common atroventricular canal (D'Cruz, Lendrum, and Novak, 1964), single coronary artery (Campeau, Ruble, and Cooksey, 1957), and pulmonary artery aneurysm and atrial septal defect (Zajtchuk, Gonzalez-Lavin, and Replogle, 1973) have been described. Coarctation of the pulmonary trunk (Sondergaard, 1954; Smith et al., 1959; Thrower, Abelmann, and Harken, 1960) has also been described with a variety of congenital

<table>
<thead>
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<th>TABLE</th>
<th>Summary of haemodynamic data</th>
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<tr>
<th>No.</th>
<th>Date</th>
<th>Pulmonary artery beyond coarctation</th>
<th>Right ventricular outflow</th>
<th>Right atrium</th>
<th>Pulmonary systemic flow ratio</th>
<th>Gradient across the pulmonary artery coarctation</th>
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<tbody>
<tr>
<td>Preop.</td>
<td></td>
<td>Pressure (s/d) (kPa)</td>
<td>Oxygen sat. (%)</td>
<td>Pressure (systolic) (kPa)</td>
<td>Oxygen sat. (%)</td>
<td>Pressure (mean) (kPa)</td>
</tr>
<tr>
<td>1</td>
<td>Feb. '59</td>
<td>8/3.3</td>
<td>82</td>
<td>12.0</td>
<td>81</td>
<td>0.7</td>
</tr>
<tr>
<td>2</td>
<td>Feb. '74</td>
<td>4.4/2.0</td>
<td>81</td>
<td>15.3</td>
<td>76</td>
<td>2.1</td>
</tr>
<tr>
<td>Post-op.</td>
<td>Aug. '74</td>
<td>2.5/1.2</td>
<td>60</td>
<td>6.8</td>
<td>59</td>
<td>2.0</td>
</tr>
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Conversion factor from SI units to Traditional units: 1 kPa=7.5 mmHg.
defects (Hudson, 1965). To our knowledge, however, the association of a coarctation of the pulmonary trunk with complete absence of the pulmonary valve and a ventricular septal defect has not been reported before.

The diagnosis of the precise pathology in our case was made difficult because the part of the pulmonary trunk below the coarctation and the right ventricle was acting as a single haemodynamic chamber. However, the absence of the pulmonary valve closure sound, the high location of the ejection systolic murmur, the loud early diastolic murmur in the axillae and interscapular area, in the presence of a normal pulse pressure, clinical and electrocardiographic evidence of right ventricular hypertrophy, and radiological evidence of pulmonary plethora, suggested a left-to-right shunt and pulmonary regurgitation. Withdrawal from the distal pulmonary artery branches revealed discrete stenosis, and this was thought to be at the pulmonary trunk level since the main pulmonary artery was not prominent in the lateral chest x-ray while both the pulmonary artery branches were enormously dilated.

Pulmonary regurgitation with a ventricular septal defect has a poor prognosis and most of the patients die in infancy or early childhood because of congestive cardiac failure (Gasul et al., 1966; Onesti and Harned, 1958; Venables, 1962; D’Cruz et al., 1964) or because of compression of the bronchial tree (Miller et al., 1962; D’Cruz et al., 1964). Experimental production of pulmonary regurgitation in dogs was reported to be well tolerated (Shaw et al., 1943; Barger, Roe, and Richardson, 1952), but later workers have concluded that the changes in the cardiac capacity are related to the extent of regurgitation and that in the absence of the pulmonary valve gross right ventricular enlargement and decompensation sets in rapidly (Kay and Thomas, 1954; Ratcliffe et al., 1957; Fowler and Duchesne, 1958).

The clinical status of the patient depends predominantly on the amount of left-to-right shunt and the degree of pulmonary regurgitation. In the absence of obstruction at the level of the infundibular or pulmonary trunk, severe cardiac decompensation results from systolic and diastolic overload of the right ventricle. In view of this, Venables (1962) has made a strong plea for pulmonary artery banding in these cases as a palliative procedure both to reduce the left-to-right shunt and to limit the regurgitant flow.

We believe that the long-term survival with minimal symptoms in our patient resulted from the coexisting pulmonary trunk coarctation which restricted the left-to-right shunt via the ventricular septal defect and thus reduced the pulmonary blood flow.

References


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