Pulmonary Atresia with Ventricular Septal Defect: Report of the Oldest Known Surviving Case*

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The case of a 54-year-old housewife, oldest published survivor with pulmonary atresia and a ventricular septal defect (pseudotruncus arteriosus), is reported. Her remarkably favorable course is likely related to the absence of progressive hemodynamic changes, with moderate pulmonary flow adequate for nearly normal arterial oxygenation yet without increasing pulmonary vascular resistance. This case reemphasizes the relationship of longevity to pulmonary blood flow volume with this defect.

Pulmonary atresia with ventricular septal defect (pseudotruncus arteriosus) is developmentally an extreme variant of tetralogy of Fallot.† The pulmonary circuit is perfused by a patent ductus arteriosus or collateral vessels from bronchial or other mediastinal arteries. Physiologically, this is identical to type 4 truncus arteriosus.‡ There has been no clear agreement from the embryologic point of view whether these two abnormalities should be considered separately. Anatomically, however, they differ. In the former, distal pulmonary arteries usually exist; in the latter, pulmonary perfusion is exclusively by bronchial collaterals. § Clinically they are often indistinguishable.

Survival with this defect into adulthood is rare. Most die in infancy. The oldest previously reported survivor was 43 at the time of the report.³ The purpose of this communication is to report a 54-year-old woman who has not only survived to this age but has had a very benign clinical course.

Case Report

This 54-year-old housewife (now 55) has been observed by one of the authors (ALF) since 1956. Born September 27, 1919, she was the product of a normal pregnancy and delivery. There was no known maternal rubella. Early development was normal. She was examined rarely in childhood and a murmur was first noted when she started school. At some time during the school years she first noted mild cyanosis, especially in the cold. There was never any squatting. Other than tiring more easily than her peers and experiencing dyspnea on marked exertion, she had no cardiac symptoms until 1972. She had one uneventful pregnancy and delivery at age 19.

On examination in 1956 there was slight cyanosis and clubbing, equal in the fingers and toes, and the mucous membranes were suffused. The pulse was 80 and regular. The blood pressure was 150/70 mm Hg in the right arm and 160/80 mm Hg in the right leg. The jugular venous pressure was within normal limits. The lungs were clear. A parasternal lift, near the xiphoid, was noticed. There was a systolic thrill at the second right intercostal space along the right sternal border. The heart sounds were normal except for accentuation of the single second sound. A grade 2-4/6 continuous murmur was widely heard over the anterior and posterior chest and was maximal over the second left intercostal space. The murmur was louder during systole. Routine laboratory examinations were within normal limits except for a hemoglobin of 17.8 gm.

Electrocardiogram was unremarkable. Chest radiography (Fig 1) and fluoroscopy showed a mildly enlarged heart with a prominent right ventricle. The left ventricle appeared elevated but not enlarged. The ascending aorta was markedly enlarged and calcification was present in the arch. The main pulmonary artery segment was concave. The hilar vessels were deficient but the pulmonary vascular markings were somewhat exaggerated. Rib notching was not present.

Right heart catheterization and a right atrial angiogram were carried out in February, 1957. The catheter repeatedly entered the aorta from the right ventricle but never entered a pulmonary artery. There was a 2.4 volume percent oxygen step up from the right atrium to the right ventricular outflow tract. Systemic oxygen saturation was 88 percent. Right ventricular systolic pressure was equal to systemic pressure; the right ventricular end diastolic pressure was 5 mm Hg.

![Figure 1. Erect posteroanterior chest x-ray film obtained in 1957. The heart is mildly enlarged. The elevated apex is due to right ventricular enlargement. The ascending aorta is markedly enlarged. Calcification is present in the arch. The main pulmonary artery segment is concave. While the hilar vessels are deficient, the hypertrophied bronchial collaterals simulate tortuous pulmonary arteries. The peripheral vascular markings are reticular and fairly prominent. Rib notching is not present.]
FIGURE 2. Erect lateral and posteroanterior roentgenograms obtained Dec. 1972. There is virtually no change from the 1957 radiogram (Fig 1). Right ventricular enlargement can be appreciated on the lateral view.

The right atrial cineangiogram showed filling of a very enlarged ascending aorta from the right ventricle. The main trunk of the pulmonary artery was not visualized, but when the aorta was filled, the distal pulmonary vessels were filled with contrast medium of similar concentration. There was a bidirectional, but net left-to-right shunt. Assuming that pulmonary venous blood was 97 percent saturated and that pulmonary arterial blood was equal to that in the aorta, the pulmonary-to-systemic flow ratio was 1.7/1.0.

The patient’s course was unchanged until June, 1972 when she developed lower abdominal pain following institution of estrogen therapy (conjugated equine estrogens, 1.25 mg every day) for postmenopausal symptoms. Her local physician found that her heart had enlarged and that the liver was palpable four fingerbreadths below the costal margin with hepatojugular reflux. These symptoms cleared when estrogens were stopped and a low sodium diet and digitalis were begun.

On repeat evaluation, the examination was unchanged from 1957. There were no signs of right ventricular failure. Routine laboratory examinations were within normal limits except for a packed red cell volume of 55.6 percent and a hemoglobin of 18.5 gm. The chest radiogram (Fig 2) also was unchanged. The electrocardiogram (Fig 3) showed nonspecific ST and T wave abnormalities.

Repeat cardiac catheterization, right ventriculogram and a retrograde aortogram in December, 1972 showed findings similar to the prior study. A 2.6 volume percent oxygen step up was present between the right atrium and the right ventricular outflow tract. Systemic oxygen saturation was 81 percent. The pulmonary artery could not be entered. Right ventricular, left ventricular, and systemic-systolic pressures were equal. However, the right ventricular end-diastolic pressure was elevated to 11 mm Hg. The left ventricular end-diastolic pressure was normal. There was a bidirectional, but net left-to-right shunt. Assuming that pulmonary venous saturation was 97 percent and that pulmonary arterial saturation was equal to that in the aorta, pulmonary to systemic flow ratio was 2.3/1.0.

The aortogram (Fig 4) showed a very large aorta. No pulmonary vessels arose from the ascending aorta; they arose from collaterals having their origin at the level of the ligamentum arteriosum. There was no recognizable retrograde filling of the main pulmonary artery. The right ventriculogram did not reveal a distinguishable infundibulum.
FIGURE 4. Aortogram (left) early in the injection shows a very large ascending aorta which does not give rise to pulmonary vessels. Later (right) in the injection, multiple collateral vessels can be seen arising at the level of the ligamentum arteriosum. Neither a main, left, nor right pulmonary artery can be identified.

**DISCUSSION**

The clinical, radiologic, and hemodynamic differentiation of pulmonary atresia with a ventricular septal defect (pseudotruncus) from type 4 truncus arteriosus is difficult or impossible.

Kirklin and Karp define pulmonary atresia with ventricular septal defect as a congenital malformation with the following defects:

1. No direct communication from the right ventricle to the pulmonary arteries.
2. Connection of the ventricles to a single arterial trunk whose semilunar valves have continuity with the anterior leaflet of the mitral valve.
3. Large ventricular septal defect located immediately beneath the arterial trunk.
4. Varying degree of underdevelopment of the infundibulum and right ventricle.

Collett and Edwards define type 4 truncus arteriosus as the lesion in those patients with a persistent truncus in whom neither right nor left pulmonary arteries nor a pulmonary trunk is present and in whom the lungs are supplied solely by bronchial arteries. The Van Praaghs, in their revision of this classification, preferred to consider this a solitary aorta with absence of the pulmonary artery and its branches.

If angiocardiographic studies demonstrate retrograde filling of the main pulmonary artery, this favors pulmonary atresia with ventricular septal defect over type 4 truncus arteriosus. It has been suggested by Gleason et al that right ventriculography may serve to distinguish the two. In their experience, patients with pulmonary atresia had a demonstrable, although blind infundibulum and patients with type 4 truncus arteriosus had no demonstrable infundibulum. Since some patients with distal pulmonary vessels may benefit from recently developed corrective surgical technique, this differentiation, when possible, may become increasingly important. We are uncertain into which category to place this patient, although the weight of evidence favors a solitary aorta with total pulmonary atresia (type 4 truncus arteriosus). In view of this difficulty in the present and in prior case reports of these disorders, we have not distinguished between them in comparing the longevity of this particular patient with similar ones in the literature.

Survival with this defect is related to the degree of systemic arterial oxygenation and the pulmonary blood flow. If oxygenation is insufficient, tissue hypoxia, erythemia and its resultant complications lead to an early death. Even if the blood flow initially is sufficient to maintain oxygenation, a subsequent increase in pulmonary vascular resistance may result in a progressive decrease in pulmonary blood flow. In their recent review of the literature, Garcia et al found only five examples of patients with this defect surviving into the third decade, in addition to their case. All six of these patients had large bronchial arteries and a relatively large volume of systemic-pulmonary collateral blood flow. This was also the case with our patient. Her particularly benign course, allowing her to become the oldest known survivor with this defect, is presumably related to her moderate pulmonary blood flow, which has been adequate to maintain peripheral arterial saturation at only slightly reduced levels, yet not productive of a progressive rise in pulmonary vascular resistance. Surgical therapy was not advised because her course had been so benign, and neither a totally corrective procedure (closure of the ventriculo-septal defect and anastomosis of the right ventricle to the pulmonary arteries by an aortic homograft) nor a palliative one (pulmonary artery banding) was feasible due to absence of demonstrable distal pulmonary arteries.

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**REFERENCES**

A New Plastic Operation for Pectus Excavatum: Sternal Turnover Surgical Procedure with Preserved Internal Mammary Vessels*

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The so-called turnover operation for pectus excavatum in older age groups has been disappointing due to postoperative complication such as fistula formation with necrosis of bones and muscles. Thus, the need of preserved vascular pedicle to the sternum was stressed in this type of operation. A new sternal crossover operation keeping the bilateral internal mammary vessels preserved was developed and satisfactorily applied on three clinical cases. The results indicate that this method is a useful sternal crossover procedure for patients over 15 years of age.

Surgical treatment of funnel chest began with the report of Ludwig Meyer in 1911. Thereafter so many reports appeared on the surgical procedures involved, that space does not permit elaboration on them, but there seems to be none which can be called decisive or established. One of the recent noteworthy surgical procedures without the use of splint is the so-called sternal turnover operation developed by Wada et al.5 The authors have made a number of trials with this method during the last several years, but in patients over 15 years of age there was a higher incidence of unfavorable postoperative complications; necrosis of bones and muscles, and fistula formation developed, much affecting postoperative motility. The incidence of these kinds of complication in our consecutive series of 26 patients older than 15, was 46 percent. With this in mind, the authors developed a method of performing sternal turnover operation with blood flow of the bilateral internal mammary vessels kept intact. This procedure was performed on three cases and the postoperative course of more than three months has been satisfactory in each case. A report is presented with a recommendation that this procedure be employed on patients over 15 years of age.

Clinical Material

Table 1 shows the three clinical cases aged 7, 23 and 29, all with severe pectus excavatum. Fluid retention volume in the funnel cavity was 70 ml, 120 ml and 130 ml respectively, indicating the severity of this entity. Two were male and one female and the surgical results were all good with primary healing in all cases. No complication was observed in any of the cases.

Selection of funnel chest patients for operation is based upon the following criteria:
1. Obvious, severe sternal depression.
2. Marked displacement of the heart.
3. Radiographic evidence of compression of the depressed sternum on the heart.
4. Cardiac complaints were not considered, but the possibility of chest deformity creating a psychologic burden on the patient in future and present life was regarded as one of the conditions.

Case 1 is the first case in which this surgical procedure was performed to evaluate whether the method was technically

Table 1—Clinical Materials and Surgical Results

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age</th>
<th>Sex</th>
<th>Severity of pectus excavatum</th>
<th>Combined anomalies</th>
<th>Surgical results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7</td>
<td>male</td>
<td>severe (70ml)*</td>
<td>none</td>
<td>primary healing</td>
</tr>
<tr>
<td>2</td>
<td>23</td>
<td>male</td>
<td>severe (150ml)*</td>
<td>VSD</td>
<td>primary healing</td>
</tr>
<tr>
<td>3</td>
<td>29</td>
<td>female</td>
<td>severe (160ml)*</td>
<td>none</td>
<td>primary healing</td>
</tr>
</tbody>
</table>

*Volume of water retained in the funnel cavity

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Figure 1. Schematic depiction of the surgical procedure.