Congenital Absence of the Pulmonary Valve with Atrial Septal Defect Surgically Corrected with Aortic Homograft

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A 20-year-old man with congenital absence of the pulmonary valve, secundum atrial septal defect and partial atrioventricular block is described. This combination of lesions has not been reported previously. Hemodynamic studies revealed left-to-right atrial shunt and low pulmonary artery pressures presumably caused by increased compliance of the aneurysmal pulmonary trunk. Corrective surgery, consisting of closure of two atrial septal defects and aortic valve homograft replacement, was successfully done.

Congenital absence of the pulmonary valve is a rare anomaly. To date, approximately 57 proved cases have been reported in the English literature. It is generally accompanied by other cardiac anomalies, the most common being ventricular septal defect with or without infundibular stenosis.1-5 Its occurrence as an isolated anomaly has been described only five times previously.1-5,8

This paper reports a case with congenital absence of the pulmonary valve and secundum atrial septal defect. This combination of lesions has not been previously described. Open-heart surgery consisting of closure of the septal defect and aortic valve homograft replacement of the absent pulmonary valve was successfully done.

CASE REPORT

The patient, a 20-year-old black man, was referred from another hospital on November 24, 1969 for evaluation of a heart murmur discovered during a routine preemployment physical examination. At age three years, he was found to have an abnormal heart. However, he has been asymptomatic and has participated in active sports. Physical examination revealed a well-nourished, well-developed, acyanotic man in no distress. Pulse was 60 per minute, respiration 20 per minute, and blood pressure 100/80 mm Hg. The radial and femoral arterial pulses were normal, and no abnormal arterial and venous pulsations were noted in the neck. Examination of the heart revealed precordial bulge, moderate parasternal heave, and systolic pulsations at the second and third left interspaces parasternally. A systolic thrill was present at the same area. S1 was normal; S2 was single and of normal intensity at the left base. An ejection systolic click, showing expiratory accentuation, was present along the left sternal border.

A chest roentgenogram before surgery (left), and six months after surgery (right). Note prominent pulmonary artery segment in both films, and the reduction in heart size after surgery.
CONGENITAL ABSENCE OF PULMONARY VALVE

The auscultatory findings. The single S2 was identified to be the aortic component, and was followed by the diastolic murmur after an interval of 40 milliseconds. No definite pulmonic closure sound was observed. The chest roentgenograms obtained in the frontal (Fig 1), lateral and oblique projections revealed marked prominence of the pulmonary artery segment which was noted to pulsate vigorously during fluoroscopy. In addition, right atrial and right ventricular enlargement was present. The pulmonary vascular markings appeared normal. The electrocardiogram and vectorcardiogram (Fig 2) showed frontal QRS axis of +100°, PR interval of 0.28 sec, QRS duration of 0.11 sec, and right bundle branch block with right ventricular hypertrophy. Another tracing obtained a few days previously showed intermittent second degree, Mobitz type I (Wenckebach) A-V block. A diagnosis of congenital absence of the pulmonary valve was made.

Cardiac catheterization performed on November 26, 1969 demonstrated a left-to-right shunt localized by hydrogen inhalation curves and by angiocardiography to be at the atrial level. The right ventricular systolic pressure was low, and identical end-diastolic pressures of the right ventricle and pulmonary artery were observed. The electrocardiogram at this time revealed first degree A-V block, and His bundle electrogams were obtained. Following intravenous atropine and isoproterenol (Isuprel) infusion, shortening of the PH but not of the HQ intervals was observed (Table 1). Cineangiocardiography, with injection of contrast medium into the pulmonary trunk, demonstrated gross pulmonary regurgitation. The pulmonary trunk and main branches were markedly dilated, and the right ventricle was moderately enlarged. Immediate opacification of a large right atrium from a normal-sized left atrium, across a suspected atrial septal defect, was noted. The left ventricle and aorta appeared normal.

Open-heart surgery with the aid of cardiopulmonary bypass was done on May 5, 1970. Two secundum atrial septal defects measuring 1.5 x 1.0 cm and 0.5 x 0.5 cm, respectively, were closed by direct suture. The pulmonary trunk was aneurysmal and thin-walled, and had a maximal diameter of 7.5 cm (Fig 3). On opening this vessel, only rudimentary valve-like tissues were noted. The proximal portion of the pulmonary trunk was resected, and a 2.4 cm diameter aortic homograft valve was sutured to the annulus and to the distal border. This was followed by a grade 3/6 coarse ejection systolic murmur, showing peak intensity before midystole and terminating before S2, at the second and third left interspaces. An early diastolic murmur of similar frequency and intensity was also present along the left parasternal area, starting a brief interval after S2 and lasting through late diastole. The rest of the physical findings were normal.

A phonocardiogram and carotid pulse tracing confirmed

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<tr>
<th>Table 1—Cardiac Catheterization Data November 26, 1969 of Patient, 20 Years Old, BSA 1.7 M²</th>
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<tbody>
<tr>
<td>Right Heart Catheterization</td>
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<td>SA</td>
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Blood flow (L/Min/M²): Pulmonary = 5.0 Systemic = 2.8 Qₚ/Qₛ = 1.8
Peripheral vascular resistance (dynes-sec-cm⁻²/M²BSA): Pulmonary = 64 Systemic = 1586 Rₛ/Rₚ = 0.04
FIGURE 3. Findings at surgery. (A) The aneurysmal thin pulmonary trunk opened. (B) The exposed rudimentary pulmonary valve consisting mostly of knobbings of endocardial tissue (arrows). (C) The aortic valve homograft in place. RV — right ventricle; PT — pulmonary trunk; AV — aortic valve homograft.

pulmonary artery. Pressures (mm Hg) obtained postoperatively were: right ventricle 47/8; pulmonary artery 17/11; and left ventricle 97/9. Cardiac output obtained by an aortic electromagnetic flow probe was 4.4 L/min/M².

Hemodynamic studies using a Carolina electromagnetic flowmeter and a high fidelity catheter-tip transducer (P-866, Statham) were also done at the time of surgery. Using a vector approach for the display of ventricular function, vector loops derived from left ventricular pressure and aortic phasic flow were obtained prior to and after bypass (Fig 4). Before bypass, the pressure-flow loops revealed left ventricular stroke power, stroke work and minute work of 443 gram-meters per second, 97 gram-meters, and 9.70 kilogram-meters, respectively. Corresponding values obtained post-bypass, and after removal of the caval cannulae, were 325 gram-meters per second, 98 gram-meters, and 8.82 kilogram-meters, respectively. Vector loops derived from the ventricular pressure curve and from its first derivative, dP/dt, were also obtained before and after bypass. The ratio of \[
\frac{dP}{dt} = \frac{1P}{1P}
\]
was derived from the slope of the initial segment of the loop (during isovolumic contraction) to estimate the myocardial contractile state. Before bypass, contractility index was 87 second⁻¹, and after bypass 127 second⁻¹ (Fig 4).

The postoperative course was uneventful, and the patient was discharged 12 days after surgery. He has remained asymptomatic, and follow-up examinations have demonstrated moderate splitting of S₂ (0.05-0.07 sec), disappearance of the diastolic murmur, but persistence of systolic thrill and murmur at the upper left sternal border. Serial chest roentgenograms showed regression of heart size, although the pulmonary artery segment has remained prominent (Fig 1). No calcifications in the homograft site have been observed. The electrocardiograms during the first and second week after surgery demonstrated normal PR intervals (0.16 sec). However, subsequent tracings showed varying first degree and second degree (Wenckebach) A-V block. The vector-cardiograms also showed persistence of the right bundle branch block but slow regression of the right ventricular hypertrophy.

Cardiac catheterization was repeated on March 17, 1971. The previous left-to-right atrial shunt was no longer demonstrated by hydrogen inhalation curves and by oximetry, but a significant systolic pressure gradient was noted across the pulmonary valve. Intravascular pressures (mm Hg) were: right atrium a = 7, v = 9, m = 6; right ventricle 52/7; pulmonary artery 19/ m = 13; and brachial artery 130/78 (m = 90). Cardiac output was 5.5 L/min/M². No angiocardograms were performed.

FIGURE 4. Vector display of left ventricular function before and after surgery. Time interruptions are 4 msec, and arrows indicate direction of inscription. From the areas of the pressure-flow loops (above), stroke power, stroke work and minute work have been derived. Note steeper slope of the initial (isovolumic) limb of the pressure-dP/dt loop (below) after surgery, indicating improved contractility. SP — stroke power; SW — stroke work; MW — minute work; CI — contractility index.

In 1969, Osman and co-workers collected 40 proved cases of congenital absence of the pulmonary valve and added eight more of their own. Nine other cases have appeared in the English literature to make a total of at least 57. To our knowledge, the occurrence of atrial septal defect as the associated cardiac anomaly has not been previously described. Second degree A-V block,

DISCUSSION

In 1969, Osman and co-workers collected 40 proved cases of congenital absence of the pulmonary valve and added eight more of their own. Nine other cases have appeared in the English literature to make a total of at least 57. To our knowledge, the occurrence of atrial septal defect as the associated cardiac anomaly has not been previously described. Second degree A-V block,
observed in our case, also has not been reported. First degree A-V block has been mentioned only occasionally.\textsuperscript{18,17} Whether the partial A-V block in our case was related to the associated atrial septal defect is a matter of conjecture. Partial or complete A-V block in isolated secundum atrial septal defect is rare.

The clinical data in our case, particularly the auscultatory findings, were quite typical of absence of the pulmonary valve. The age, lack of symptoms and the normal pulmonary vascular markings in the chest roentgenograms, suggested that the lesion could have been an isolated one.

In spite of the dual mechanisms for the right ventricular volume overload, namely, the left-to-right atrial shunt and the pulmonary regurgitation, it is of interest that the pressures in the right heart chambers were not elevated. In fact, the systolic pressures in the right ventricle and pulmonary artery were low in spite of the obviously large stroke volumes. This can only be explained by increased compliance of the aneurysmal pulmonary compression chamber. The pulmonary or right ventricular hypertension in the previously reported cases must have been chiefly due to the associated large ventricular septal defects. The equalization of the late diastolic pressures of the right ventricle and pulmonary artery, observed in our case, is a well known phenomenon in gross pulmonary regurgitation.

Because of the paucity of reports on congenital absence of the pulmonary valve occurring as an isolated anomaly\textsuperscript{5,6-8} the prognosis of patients with this condition is not known. Severe heart failure has been observed in the immediate neonatal period.\textsuperscript{5,7} On the other hand, absence of the pulmonary valve was an accident autopsy finding on a 73-year-old man who had acquired aortic insufficiency from bacterial endocarditis.\textsuperscript{8}

The low pressure, low-resistance pulmonary vascular bed in our case obviously played an important role in maintaining his asymptomatic state. Nevertheless, because of the right heart enlargement and of the atrial septal defect, corrective surgery was done. The ventricular function studies at the time of surgery were confined to those of the heart since no adequate-sized flow probe for the aneurysmal pulmonary trunk was available. Right ventricular contractility indices before and after cardiopulmonary bypass, determined similarly to those of the left ventricle, would have been most informative. The hemodynamic studies at the time of surgery, and the clinical as well as repeat hemodynamic evaluations following surgery, indicate that pulmonary valve competency has been maintained. However, mild pulmonary stenosis, most likely due to narrowing of the homograft valve ring, appeared immediately following surgery and has persisted. The long-term fate of this graft cannot be predicted but there is suggestive evidence, in adults at least, that this type of valve is preferable to prosthetic ones for aortic valve replacement.\textsuperscript{16}

Aortic valve homograft for the treatment of this condition has also been reported once previously.\textsuperscript{19} We believe that it is the procedure of choice for the correction of this anomaly.

REFERENCES