CONGENITAL ABSENCE OF PULMONARY VALVE AND VENTRICULAR SEPTAL DEFECT: SURGICAL REPAIR WITH HOMOLOGOUS DURA MATER VALVULAR PROSTHESIS

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INTRODUCTION

Congenital absence of pulmonary valve was first described by Chevers in 1847,[1] and since that time, at least 150 cases have been reported. It may occur alone or in association with other congenital malformations of the heart, such as tetralogy of Fallot,[2,3] ventricular septal defect,[4] obstruction of the right ventricular outflow, and dilatation of the pulmonary arteries.[5] The most frequent coexisting lesion is a large ventricular septal defect, almost always infracristal.[6]

The characteristic pathological configuration is a continuity between the right ventricular infundibulum and the pulmonary trunk eventually limited by a hypoplastic pulmonary valve, and an aneurysmatic dilatation of the pulmonary trunk and its branches. This dilatation may assume huge proportions and compress the tracheobronchial tree, producing respiratory distress and obstructive pulmonary emphysema in the first year of life.[5,6,7] The development of congestive cardiac failure may worsen the clinical picture and induce high mortality in this group of patients.[7]

In the absence of other cardiac malformations, pulmonary insufficiency is well tolerated,[8] but in those patients with multiple cardiac defects, cardiac failure may present difficulties in clinical management. Although surgery is a serious consideration in this condition, indications for total intracardiac repair are not well established, since the natural history of this condition is poorly understood.[9]

We present a patient who had atrial and ventricular septal defects, obstruction to the right ventricular outflow, and absent pulmonary valve, which were surgically repaired. Embriologic, anatomic, and surgical considerations of the pathology are discussed, and the importance of pulmonary insufficiency is emphasized.

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PATIENT MATERIAL

Case Report

A 17-year-old male was referred to our institution with cyanosis which had been apparent since infancy. He had dyspnea on exertion, and a history of repetitive pulmonary infections.

Examination revealed normal physical development, digital clubbing, and mild cianosis of the extremities. The vital signs were normal, and the peripheral pulses were equal and of normal character. The lungs were clear to auscultation. The cardiac apex was at the 4th and 5th intercostal space at the anterior axillary line. Systolic and diastolic thrills were felt and a Grade IV/VI harsh systolic murmur was audible over the precordial surface, followed by a single second heart sound and a Grade IV/VI diastolic murmur. The electrocardiogram showed right Bundle Branch block and right atrial and ventricular hypertrophy (Fig. 1). A chest X ray revealed an enlarged heart with a markedly dilated pulmonary trunk and its branches, and decreased peripheral pulmonary vasculature (Fig. 2 A and B). A vectorcardiogram confirmed right atrial and ventricular hypertrophy, and showed a delay in activation of basal portions of the right ventricle. At cardiac catheterization, the aortic systolic pressure was 113 mm Hg with no gradient across the aortic valve. The pressure in the right ventricle was 129.0/11.5 mm Hg and in the left ventricle 133.0/15.5 mm Hg. The mean right atrial pressure was 7.5 mm Hg. The pulmonary artery pressure was 56.5/25.5 mm Hg. A bi-directional shunt at the atrial and ventricular level was shown by oxygen saturations.

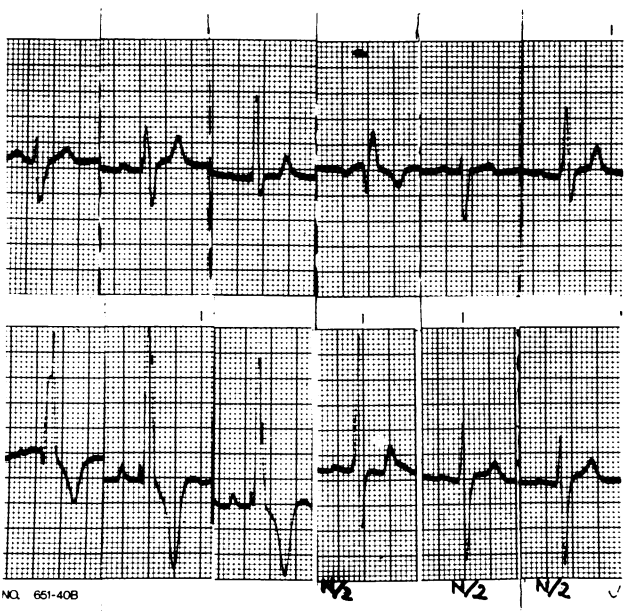


Fig. 1. Electrocardiogram showing right bundle branch block and right atrial and ventricular hypertrophy.

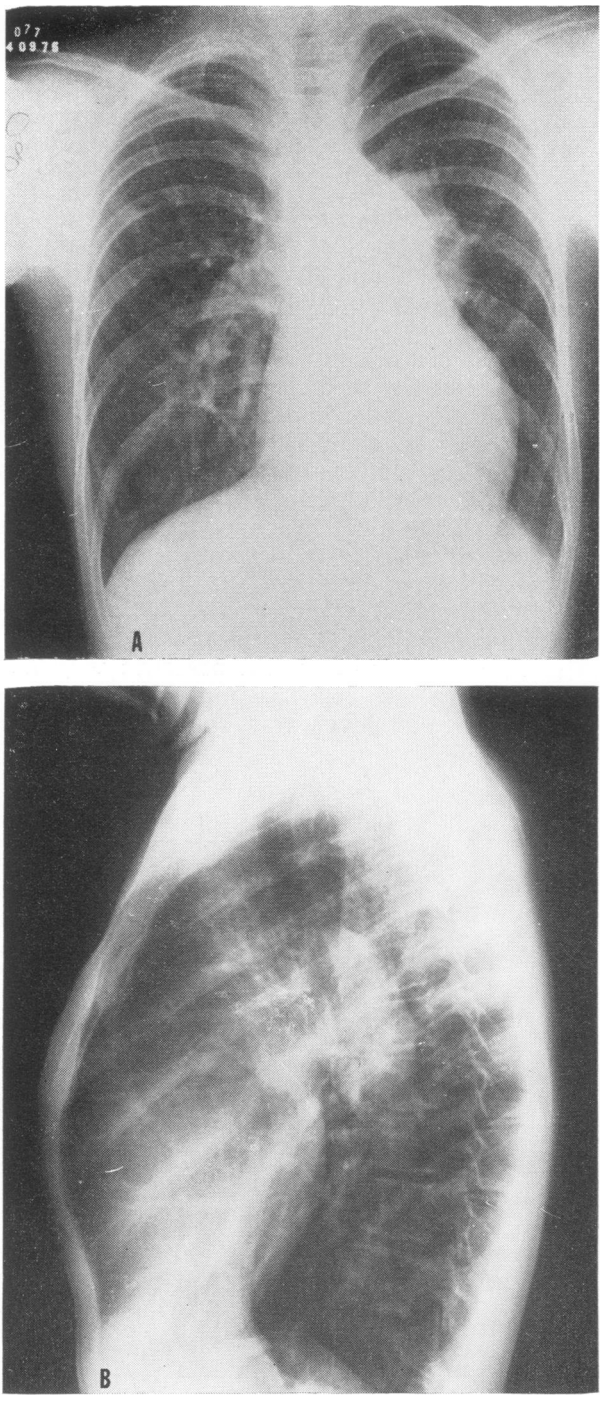
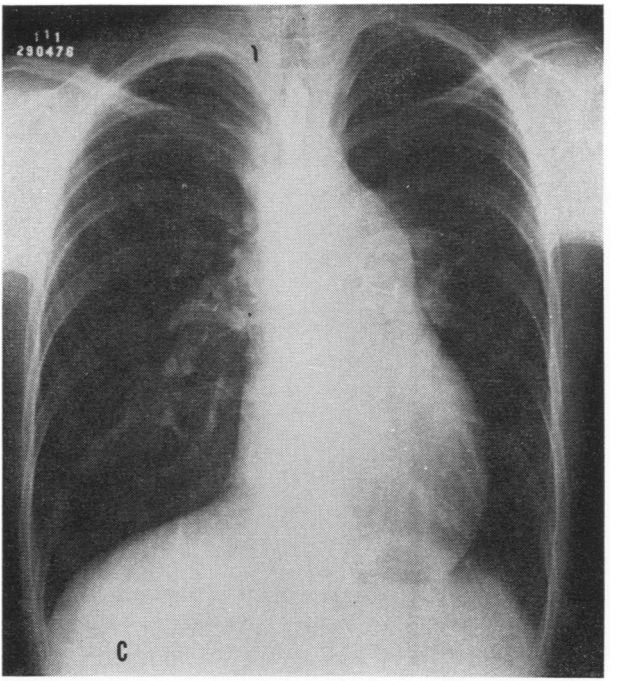


Fig. 2. A and B: Preoperative frontal and lateral Roentgenograms of the chest. Dilatation of the pulmonary artery and its branches. C and D: Postoperative frontal and lateral Roentgenograms showing the homologous duramater prosthesis in the pulmonary annulus.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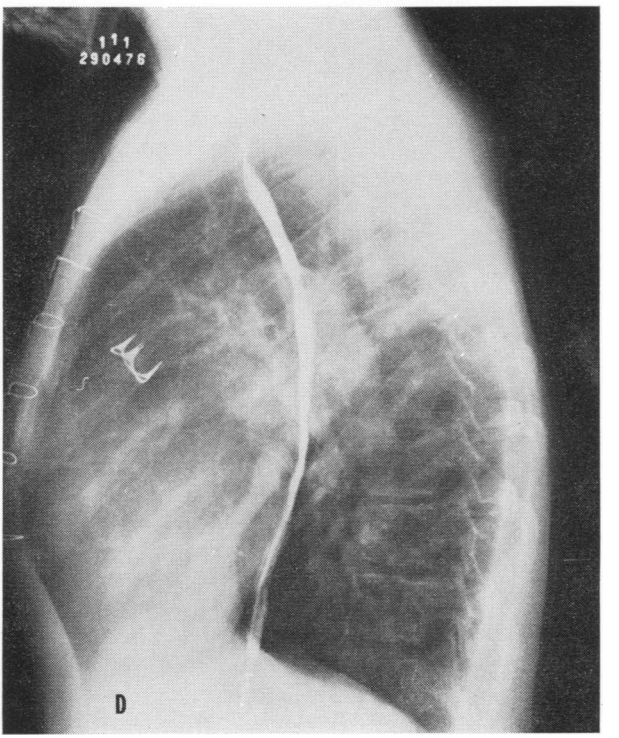
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Cineangiocardiographic study showed an enlarged right ventricle, a dilated but normally-placed pulmonary artery, and an irregular configuration at the pulmonary valve annulus. Absence of pulmonary valve leaflets was noted. Infundibular stenosis and an infracristal ventricular septal defect were also identified (Fig. 3). The left ventricle was enlarged. The ascending aorta was of normal size, and there was aortic-mitral continuity.

Surgical correction was recommended with a diagnosis of *absent pulmonary valve, ventricular septal defect* and *atrial septal defect.* Through a midline sternotomy, total cardiopulmonary bypass was established at normothermia. The right ventricle was entered through a transverse incision and an infundibulectomy was performed. An infracristal ventricular septal defect approximately 2 cm in diameter was closed with a Teflon patch fixed with continuous sutures. The pulmonary trunk was opened longitudinally and a homologous duramater valvular prosthesis[10] (20 mm internal diameter) was inserted on the valvular ring with multiple interrupted sutures. Through the right atrium, the atrial septal defect, ostium secundum type, was closed with a Teflon patch fixed with continuous sutures. After recovery of the heartbeat, the cardiopulmonary bypass was discontinued. The caval and arterial cannulae were removed and the sternotomy closure performed. After two days in the recovery room and ten days in the ward, the patient was discharged. Periodic examination (now at the 6th postoperative month) reveals improvement in the patient's physical condition with no cardiovascular symptoms. Figure 2 C and

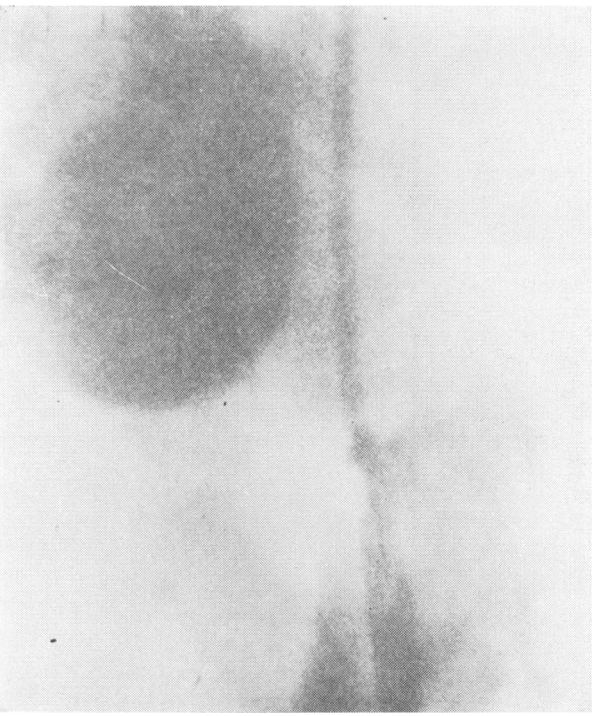

Fig. 3. Photograph of a cine frame of a right ventricular injection. Note the marked aneurysmal dilatation of the pulmonary artery and absence of pulmonary valve.

D shows a frontal and lateral Roentgenogram of the chest at the time the patient was discharged.

DISCUSSION

Absence of the pulmonary valve may be attributed to the deficient development of cusps primordia in the trunco-conal septum. When the trunco-conal septum is inadequately placed, around the 7th week of embriogenic life, ventricular septal defect and pulmonary stenosis may result.[11,12,13] Isolate absence of the pulmonary valve has been explained by intrauterine infection, but this hypothesis is based upon only two cases.[9]

The causes of aneurysmatic dilatation of the pulmonary trunk or its branches, associated with absence of the pulmonary valve with or without other cardiac malformations, are not well established. Histologic studies revealed structural malformations of the pulmonary walls, characterized by elastic fiber disruptions and absence of the media, muscular hypertrophy, or similarities with the aortic wall.[14] It is reasonable to attribute the dilatation to an association of factors, such as elevated intrauterine pulmonary vascular resistance, increased pulmonary flow in the absence of the ductus arteriosus, the presence of a ventricular septal defect, and pulmonary or subpulmonary stenosis, if present.

Although isolated absence of the pulmonary valve rarely induces symptoms in the first year of life, aneurysmatic pulmonary arteries may compress the tracheobronchial tree, resulting in significant respiratory distress. The clinical picture worsens considerably when other cardiac malformations exist. If lobar emphysema is found, resection is mandatory before the institution of other clinical or surgical treatment.

Our patient, although presenting severe cardiac malformation, had mild cardiovascular dysfunction (NYHA Class II), differing from most cases reported, where early death occurred or surgery was performed during the first years of life. The age and reasonable clinical status might be explained by a hemodynamic balance between the intracardiac lesions: infundibular stenosis with reduced pulmonary flow, and probably balanced interventricular shunt.

In this case, three fundamental factors were considered: aneurysmatic dilatation of the pulmonary artery, the intracardiac malformations, and pulmonary valvular insufficiency. Important dilatation of the pulmonary trunk was present, but no compression of the airways or pulmonary emphysema could be found on the X ray. We did not correct the aneurysmatic dilatation since we expected that the hemodynamic normalization of the pulmonary circulation after surgery could constrain further development of the aneurysm.

Two intracardiac malformations were corrected: an atrial defect and an infracristal subaortic ventricular septal defect. Infundibular resection was performed. An otherwise normal infundibulum and the correct placement of the aortic valve in relation to the intraventricular septum excluded the diagnosis of tetralogy of Fallot.[16] There is no universal agreement regarding insertion of a pulmonary valve prosthesis, but since increased

postoperative morbidity and mortality might be associated with the persistence of pulmonary insufficiency by right ventricular overload and failure[17,18], and, since this might be aggravated by the ventriculotomy, we elected to correct the pulmonary regurgitation. It is important to report that in each of seven patients in whom surgical repair of absence of pulmonary valve and ventricular septal defect was successfully performed, a homograft was used in the pulmonary valve.[4] The late prognosis may also be compromised when pulmonary insufficiency persists,[2] and we believe that it is an unnecessary surgical risk to consider later reoperation when pulmonary insufficiency can be corrected during the initial procedure.

SUMMARY

Chevers in 1847 reported the first case of congenital absence of the pulmonary valve, a relatively rare malformation, usually associated with a ventricular septal defect and right ventricular outflow obstruction.

This report presents a patient with absent pulmonary valve associated with atrial and ventricular septal defects, right ventricular outflow obstruction, and dilatation of pulmonary arteries. The pulmonary insufficiency was corrected with implantation of a homologus duramater valvular prosthesis. Follow-up data at six months revealed that the patient had improved and was free of symptoms.

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