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CONGENITAL ABSENCE OF THE PULMONARY VALVE ASSOCIATED WITH PULMONARY STENOSIS, LARGE DUCTUS ARTERIOSUS AND INTACT VENTRICULAR SEPTUM*

Case Report

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SUMMARY: Yurdakul Y, Atasoy Ş, Sarıosmanoğlu N, Özer S, Bilgiç A. (Departments of Thoracic and Cardiovascular Surgery and Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Congenital absence of the pulmonary valve associated with pulmonary stenosis, large ductus arteriosus and intact ventricular septum. Case report. Turk J Pediatr 1994; 36: 163-169.

A six-day-old neonate was diagnosed with a severe form of the syndrome of absent pulmonary valve associated with pulmonary stenosis, aneurysmal dilatation of the pulmonary artery and rare findings including an intact ventricular septum and large ductus arteriosus. The patient underwent surgical repair by closed technique. Cardiac catheterization data, hemodynamic and clinical findings, and surgical technique are reported.

Congenital absence of the pulmonary valve is a rare cardiac anomaly. An especially severe form occurs with ventricular septal defect and pulmonary stenosis. The usual findings are respiratory distress, aneurysmal dilatation of the pulmonary arteries and pulmonary stenosis. *Key words: congenital absence of pulmonary valve, intact ventricular septum, closed surgical technique.*

Absent pulmonary valve is a rare congenital heart defect usually associated with ventricular septal defect, patent ductus arteriosus, endocardial cushion defect, double outlet right ventricle and Marfan's syndrome[1,2].

Pulmonary insufficiency and massive or aneurysmal dilatation leading to tracheobronchial tree compression are the common features of absent pulmonary valve syndrome[3-5].

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In infants and neonates, absent pulmonary valve syndrome is associated with severe respiratory problems, often due to tracheobronchial tree compression, and can cause respiratory and cardiac failure. Treatment of this syndrome can include medical and surgical approaches. Surgical treatment includes aneurysmorrhaphy, aneurysmectomy, division of right pulmonary artery and tubular graft reconnection with the main pulmonary artery or anastomosis with the superior vena cava[2]. In older patients, some physicians do not recommend pulmonary valve replacement[6], while others do recommend this procedure in order to prevent pulmonary regurgitation[7].

Case Report

A six-day-old male neonate weighing 3100 g was cyanotic, tachypneic and in moderate respiratory distress. Cardiac findings included increased right ventricle impulse, loud and single S_2, and grade 2-3/6 systolic and diastolic murmur at the left upper sternal border. Chest X-ray showed mild cardiomegaly.

Echocardiography showed no ventricular septal defect, dilated pulmonary arteries, a 20 mm diameter main pulmonary artery, stricture in pulmonary annulus and no pulmonary valve (Fig. 1-a).

Cardiac catheterization data revealed 63 mm Hg pressure in both the right and left ventricles, an intact ventricular septum, aneurysmal dilatation of the main pulmonary artery, and large ductus arteriosus (Fig. 2).

The infant underwent surgical repair on the sixth day of life. Using left posterolateral thoracotomy, we reached the pericardium. After opening the pericardium 1 cm above and parallel to the left phrenic nerve, we were faced with an aneurysmal, dilated, main pulmonary artery. We proceeded by placing double oval shaped purse sutures on the main pulmonary artery. After pulmonary arteriotomy using a straight clamp, we enlarged the pulmonary annulus in two steps, so as to allow pulmonary flow and minimize bleeding. Then, we resected the pulmonary artery aneurysm using a partial occluding vascular clamp, and the arteriotomy was sutured. Patent ductus arteriosus was ligated (Figs. 3-4).

On the sixth postoperative day, the main pulmonary artery measured 9 mm in diameter by echocardiograpy (Fig. 1-b).

The patient remained symptom-free during a one-year postoperative follow-up period, and normal-sized pulmonary arteries were seen on chest X-ray (Fig. 5) and echocardiography (Fig. 1-c).

Discussion

Since Cheever's report in 1847, 306 cases of congenital absence of the pulmonary valve have been reported. Most of the patients had a ventricular septal

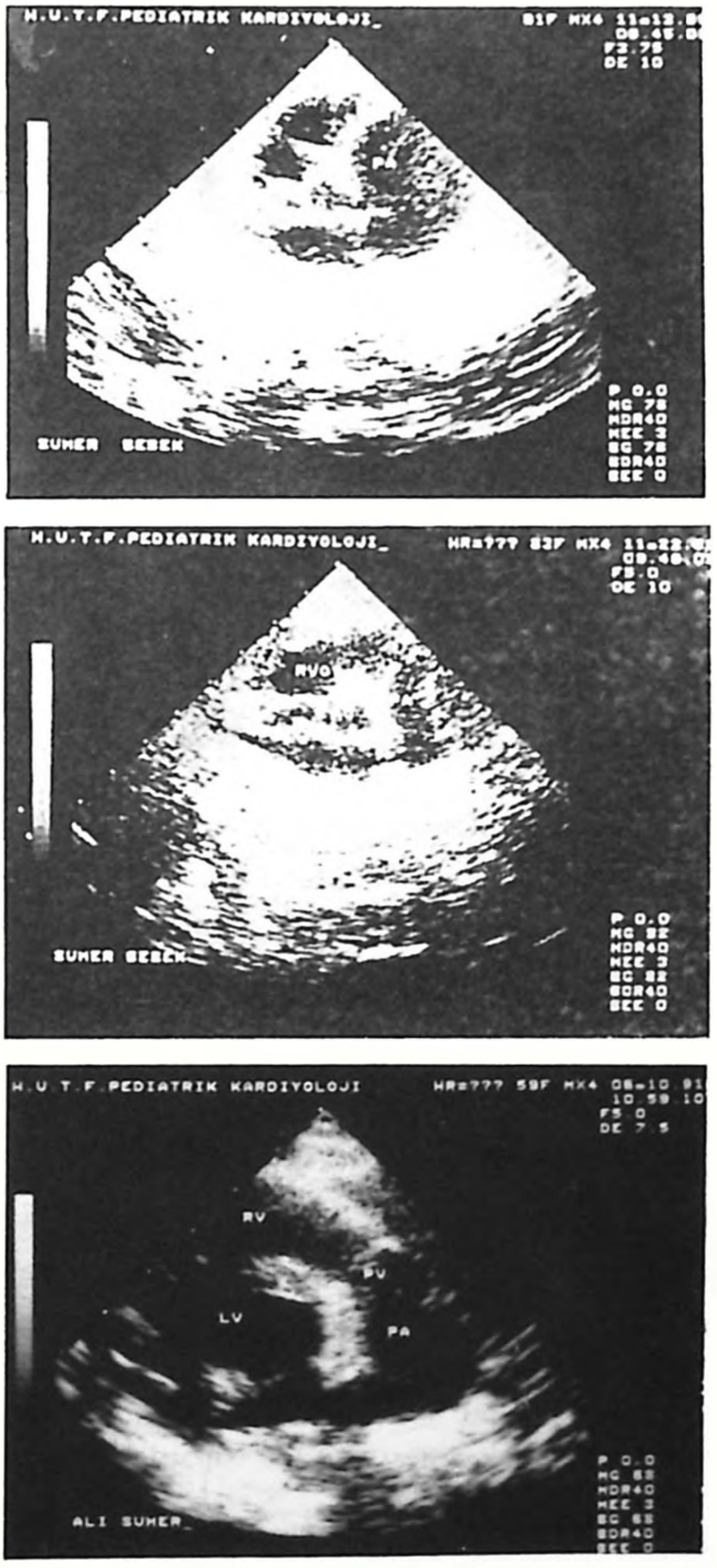

Fig. 1: Two-dimensional echocardiography (a) preoperatively showing no pulmonary valves and 20 mm main pulmonary artery, and (b) post-operatively on the sixth day showing the pulmonary artery 9 mm in diameter, and (c) after one year of follow-up showing normal pulmonary artery. (AO: Aorta, PA: Pulmonary artery, RV: Right ventricle, LV: Left ventricle, RVOT: Right ventricular outflow tract PV: Pulmonary valve annulus).

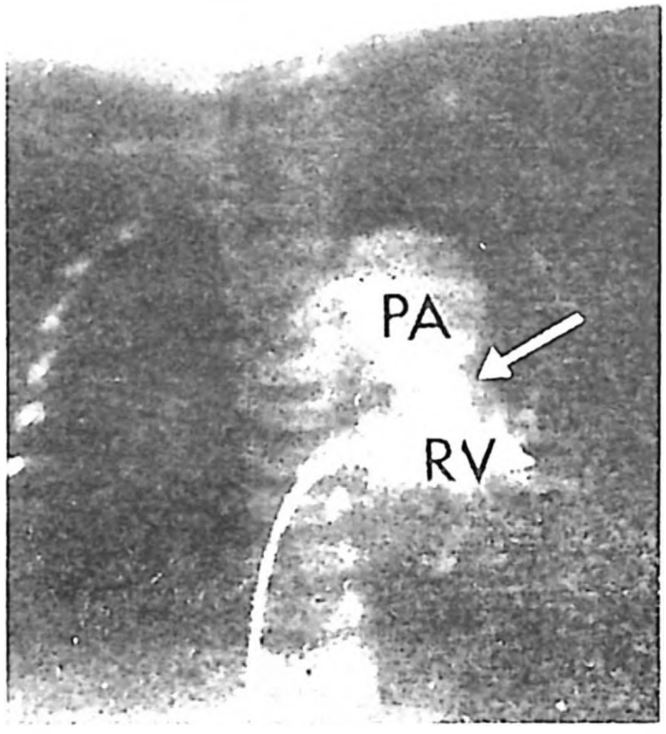

Fig. 2: Antero-posterior view of selective right ventriculogram showing aneurysmal dilatation of the main pulmonary artery. PA: Pulmonary artery, RV: Right ventricle.

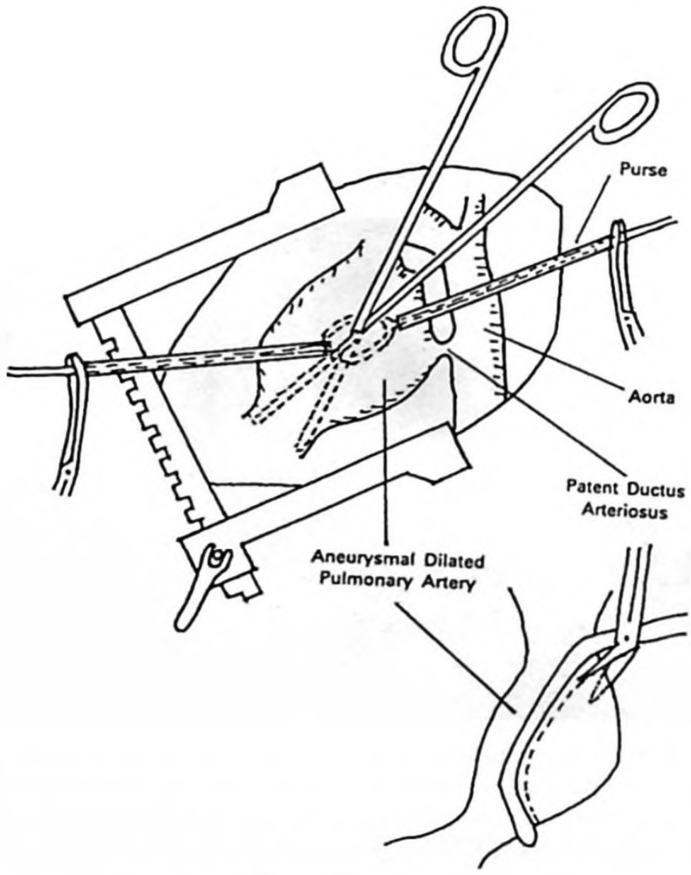

Fig. 3: Illustration of dilatation of the pulmonary annulus and resection of aneurysm.

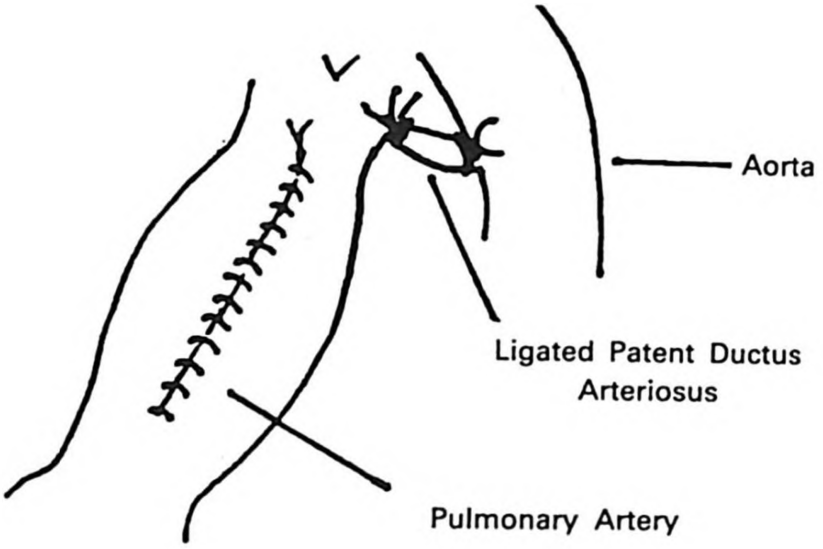

Fig. 4: Illustration showing the last image at the end of surgery.

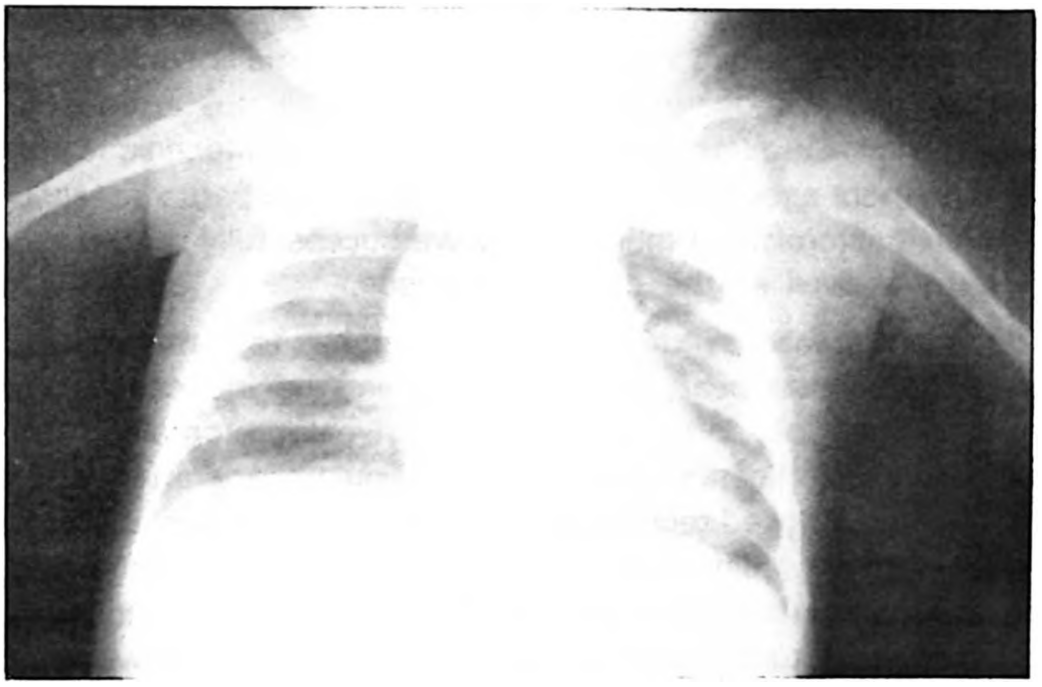

Fig. 5: Postero-anterior chest X-ray showing the normal-sized pulmonary artery.

defect, pulmonary stenosis and aneurysmal dilated pulmonary arteries. The occurrence of an intact ventricular septum with an absent pulmonary valve is very rare[1].

When the pulmonary valve is absent, severe problems arise soon after birth. Tracheobronchial compression resulting in atelectasis and obstructive emphysema cause severe respiratory distress[1].

There are several causes of aneurysmal dilatation of the pulmonary arteries, the most important of which are pulmonary stenosis and pulmonary insufficiency.

Pulmonary insufficiency increases the right ventricular stroke volume which, coupled with pulmonary stenosis results in pulmonary dilatation[3,5]. However, if the degree of stenosis is too severe, blood flow cannot produce pulmonary dilatation[8].

Patients born with absent pulmonary valves can be divided into two groups: patients in group I present with severe cardiorespiratory distress in infancy, while patients in group II display mild symptoms in infancy[3]. Our patient was in the first group.

The most important aspects of surgical management are: (1) reducing the pulmonary artery aneurysmal dilatation, (2) relieving pulmonary stenosis, and (3) performing intracardiac repair if any intracardiac anomaly is present[3,6].

A surgical approach can involve open or closed heart surgery. In open heart surgery, total correction can be achieved, while in closed technique a palliation can usually be done (e.g., pulmonary artery binding, or Blalock-Taussig shunt) depending on the cardiac anomaly.

In our case a closed technique was chosen because of the absence of intracardiac anomaly other than pulmonary stenosis and patent ductus arteriosus. In open heart surgery, a midline sternotomy is not so effective a technique for reaching the pulmonary artery aneurysmal dilatation to perform a partial resection. In this case, using left posterolateral thoracotomy we successfully reached the pulmonary aneurysm as well as the ductus arteriosus.

In absent pulmonary valve syndrome, the surgeon has to think carefully whether or not to perform open heart surgery to correct the intracardiac anomaly, if present, whether to use aortic valve homograft or heterograft to prevent the pulmonary artery insufficency and secondary pulmonary artery dilatation, or whether to employ a closed technique according to the present cardiac anomaly. Reports showed that a satisfactory result can be obtained by using an aortic valve homograft or heterograft in older patients[5-7]. A palliative operation can be used with infants to facilitate the normal development of their airways.

In case the reported patient may face residual pulmonary stenosis in the future, but we believe that elective total repair of pulmonary stenosis and insufficiency later will result in a lower risk of mortality[4].

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