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LETTER TO EDITOR

INTERMEDIATE TYPE PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM*

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Pulmonary atresia (PA) with intact ventricular septum (IVS) is a rare and life-treathening congenital anomaly which causes mortality and morbidity in infancy[1]. We report herein an infant who had PA with IVS.

A five-month-old boy was hospitalized with the signs of cyanosis and severe heart failure in October 1994. His initial diagnosis was atrial septal defect (ASD) and pulmonary stenosis (PS). Because of severe bradycardia and cardiac instability during cardiac catheterization, the procedure was not able to be performed. He was then taken to surgery urgently with ASD plus PS plus functional tricuspid insufficiency, which was diagnosed by echocardiography.

We performed the operation by the standard means of cardiopulmonary bypass using moderate hypothermia. Initially we opened the main pulmonary artery, but we could not find any orifice. Instead of the pulmonary valve, there was fibromuscular tissue (Fig. 1). We extended our incision down to the right ventricular outflow tract, resected the fibromuscular tissue and inserted a cross-annular patch. Then we closed the ASD with a dacron patch. No extra surgical procedure was considered for tricuspid insufficiency because it was not important hemodynamically. At last, the patient was taken to the intensive care unit with no inotropic support.

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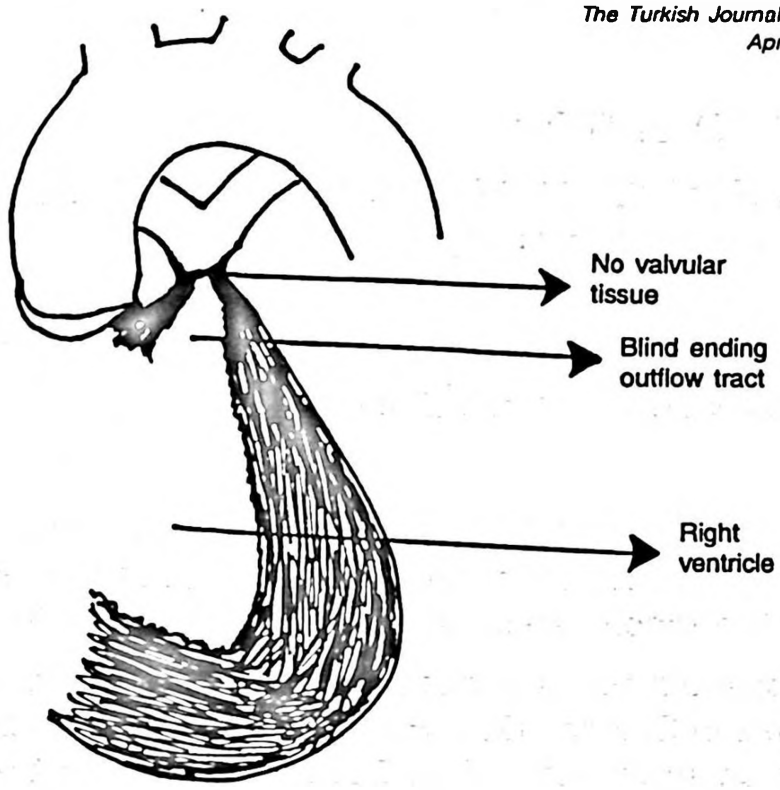

Fig. 1: Schematic drawing of the patient's right ventricular outflow tract. The base of the pulmonary trunk itself is connected to the ventricular mass, but the cavity of the trunk is separated from the ventricle by fibromuscular tissue.

Unfortunately, in the early postoperative period the patient suffered from oliguria which progressed to anuria. Peritoneal dialysis was established, but the patient died in the early postoperative period because of a cardiac arrest.

Pulmonary atresia is a congenital aberration in which there is complete blockage of the pathway from the pulmonary trunk to the ventricular mass[2]. The atresia can take one of three forms –valvular or membranous atresia, muscular atresia, and solitary arterial trunk– the differences being of considerable surgical significance[3, 4]. Also, intermediate types may be found in some cases. In such patients, the decision as to the most suitable procedure may be difficult to make[5].

Our case was an intermediate form of PA with IVS between Types I and II. We decided to continue corrective surgery because the patient had already been taken for open-heart surgery when the exact diagnosis was established.

The preoperative, definitive diagnosis gains importance especially in the intermediate and complex forms of PA with IVS[6, 7]. In order to determine the most suitable treatment, all laboratory studies, invasive (when needed) and noninvasive diagnostic tools must be performed[8].

An angiocardiography needed to have been performed in this case in order to make a definitive diagnosis and change the type of intervention. In this case, if the preoperative diagnosis had been PA with IVS, a palliative procedure could have been our choice of procedure, where a staged repair could have decreased the mortality and morbidity of this child.

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