SURGICAL CLOSURE OF AORTOPULMONARY WINDOW IN A 5 MONTHS OLD INFANT

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Aortopulmonary window is a rare congenital cardiac anomaly with serious hemodynamic consequences which requires urgent surgical treatment. Five months old girl infant who had signs and symptoms of congestive heart failure underwent surgical treatment with the diagnosis of Type I aortopulmonary window and additional fossa ovalis type atrial septal defect. Aortopulmonary window was closed through transaortic approach and by primer suturation. Fossa ovalis type atrial septal defect was closed by primer suturation. Postoperative course of the patient was uneventful.

Key words: Aortopulmonary window, surgical treatment, primer suturation

Aortopulmonary window (APW) (aortopulmonary septal defect, partial truncus arteriosus) is a rare congenital heart anomaly with an incidence of 0.1- 0.2% among the patients with congenital heart disease (1,2). The defect is caused by incomplete formation of the septum in the developing truncus arteriosus during embryogenesis which results in abnormal communication between the aorta and pulmonary artery distal to the semilunar valves. According to the classification of Mori et al., Type I defects has a communication between the ascending aorta and main pulmonary artery just above the sinus of Valsalva, Type II defects are localized between the ascending aorta and the junction of the right and main pulmonary artery, and Type III defects are a combination of these two types beginning just above the semilunar valves and reaching to the pulmonary bifurcation and proximal segment of the right pulmonary artery (3). Additional cardiac anomalies coexist with APW in 5 to 10% of the patients (4). Spontaneous closure has not been reported and important pulmonary vascular diseases develop in early life. There is a limited number of reported cases of surgical closure under 1 year of age. We report a case a 5 months old infant with Type I APW and additional atrial septal defect (ASD). Both the APW and ASD were successfully closed with direct suture closure using cardiopulmonary bypass.

CASE REPORT

The patient was a 5 months old girl infant. She was a term procust of an uncomplicated pregnancy. She had repeated respiratory infections in her history. On physical examination there was a continuous murmur (grade 4/6) on the left side of the sternum, and the second heart sound was accentuated. The infant was 4100 g in weight and 56 cm in length. She had signs and symptoms of mild congestive heart failure. The chest radiograph revealed an increased cardiothoracic index (70%). There were evidences of biventricular and left atrial enlargement on the electrocardiogram. Echocardiography showed the shunt between the ascending aorta and the main pulmonary artery and the diagnosis of APW was made showing the separate aortic and pulmonary valves with normal morphology and function. There was additional fossa ovalis type ASD. Cardiac catheterization and cineangiography which was performed to provide the definitive diagnosis and to identify associated cardiac anomalies, revealed rapid filling of pulmonary artery through the APW between the ascending aorta and pulmonary artery, separate aortic and pulmonary valves, normal coronary anatomy, and associated atrial septal defect (Figure 1). The ratio of pulmonary vascular resistance to systemic vascular resistance (Rp/Rs) was 0.12.

In the view of these findings, the patient was referred for operation. The operation was performed through median sternotomy, using cardiopulmonary bypass. After opening the pericardium the thrill above the APW was palpated. Limited dissection was made between the aorta and the pulmonary artery and right pulmonary artery was identified. Using mild hypothermia, hypoxic, hypothermic, cardioplegic arrest was performed. Right atriotomy was performed and the ASD with a diameter of 15 mm was closed by primer suturation using 5/0 prolene. The aorta was opened transversely at the level of APW. The defect with a diameter of 5 mm was identified. Following careful identification of the anatomy of the coronary arteries and aortic valve leaflets, the defect was closed by direct suture technique using two rows of continuous 5/0 prolene suture. There was no risk of damaging coronary ostium or aortic leaflets. The aortotomy and right atriotomy incision were closed and remainder of the operation was performed in usual manner. There was no thrill above the window area at the end of the operation. Postoperative echocardiographic examination of the patient showed no residual shunt. The patient had an uneventful postoperative course and was discharged 5 days after the surgery.

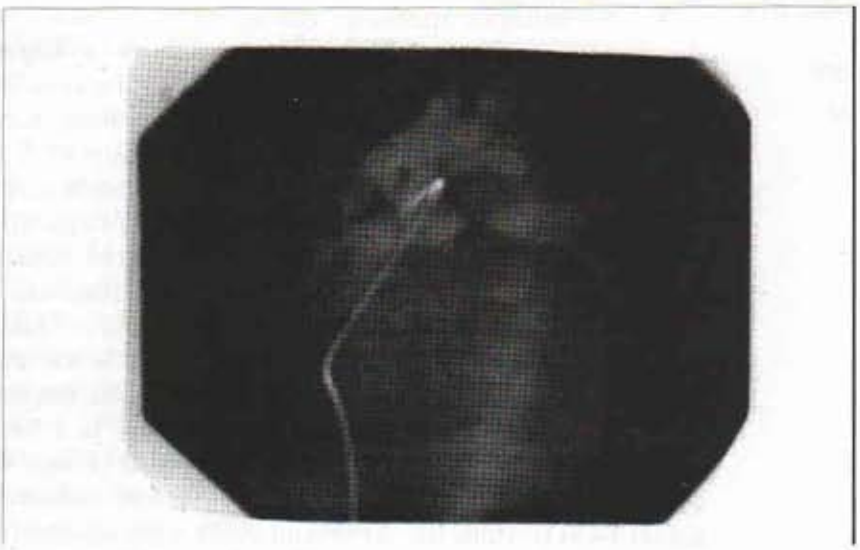

Figure 1. Cineangiographic view revealing filling of the pulmonary artery through the aortopulmonary window.

DISCUSSION

APW is a rare congenital cardiac anomaly with serious hemodynamic consequences which requires urgent surgical treatment. Since pulmonary vascular disease develops early in life in patients with APW, most of the patients die before childhood or adult life unless surgery is performed. The differential diagnosis includes patent ductus arteriosus, truncus arteriosus, ruptured aneurysm of sinus of Valsalva and ventricular septal defect with aortic incompetence.

The most common associated cardiac anomalies are patent ductus arteriosus and atrial septal defect (3). Although the diagnosis can be made by echocardiography in the majority of the cases, cardiac catheterization and cineangiography provides definitive diagnosis and identify associated anomalies. To make the definitive diagnosis, contrast material should be injected into the left ventricle and/or aortic root and the rapid filling of the pulmonary artery through window and the separate aortic and pulmonary valves must be visualized (5,6). It has been emphasized that the patients with APW have high risk of developing pulmonary vascular disease early in life and the surgery must be performed as soon as the diagnosis is made (1,3,5). The first successful surgical closure of APW was achieved by Gross in 1952 using simple ligation technique (7). In 1957, Cooley et al. reported repair of an APW with cardiopulmonary bypass and using division technique. The stenosis of the pulmonary artery and/or ascending aorta, the stenosis or injury of the left coronary artery ostium are the complications which may be encountered by using the techniques of ligation and division (3,6). With the introduction of cardiopulmonary bypass and the opportunity of deep hypothermia and total circulatory arrest when needed, direct closure of the defect under visualization became the preferred and recommended method which makes the repair easier and eliminates the risk of complications. Increased preoperative pulmonary vascular resistance ($Rp/Rs > 0.4$) is an important risk factor for mortality (1,6). Closure of the APW with a patch of Dacron or pericardium through transaortic approach is a commonly applied technique. The main advantage of this technique is not to cause any stenosis or angulation of the vessels (5,6). However, the risks of insufficient exposure, residual shunt and stenosis of the left coronary ostium by transluminal sutures are reported when the techniques of division and transaortic patch closure are used in large Type I and Type II defects (3,5). In addition there are some other risks related to the preferred patch material (Dacron or pericardium). Transpulmonary patch closure is an alternative technique but usually does not provide a sufficient exposure for Type II defects (6,7). There are some reports of successful closure of APW by hemoclip application (2), but they also report the unpleasant experience resulting in mortality (8). Although it has not been commonly reported, transaortic direct suturation is an appropriate technique for small sized APW. This technique provides complete protection of the aortic leaflets and coronary ostia. In addition, it eliminates the risks related to the patch material. In our case, the defect was small sized and we preferred direct suture closure instead of using a patch. There was not stenosis or angulation of the aorta or pulmonary artery after the closure. Coronary ostium and aortic leaflets were away from the suture area and there was no complication related to these structures.

CONCLUSION

APW is a rare congenital cardiac anomaly and associated cardiac anomalies may coexist. Preoperative increased pulmonary vascular resistance is an important risk factor for mortality. Although the surgical technique should be chosen according to the size and localization of the defect, primer suturation through aortic approach is a recommended one for small sized defects.

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