

FALLOT TETRALOGY AND SYSTEMIC TO PULMONARY ARTERY COLLATERAL CIRCULATION FROM RIGHT INTERNAL MAMMARIAN ARTERY: A VERY RARE ANATOMICAL VARIATION

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Pulmoner hipoplazi ya da atrezi ile ventriküler septal defektin birlikte olduğu hastaların pulmoner kan akımı oldukça değişkenlik gösterir. Fallot tetralojili hastalarda pulmoner hipoplazi veya atrofi halinde oldukça küçük pulmoner arterlerin mevcut olması tam düzeltme için yüksek risk grubunu oluştururlar. Bu yazıda Fallot tetralojili ve pulmoner hipoplazisi olan bir hastada farklı bir kaynaktan sağlanan kan akımı tanımlanmıştır. Sağ

internal mammarian arterden pulmoner kan akımının sağlanması oldukça nadir bir durumdur.

Anahtar kelimeler: Fallot Tetralojisi, Pulmoner hipoplazi, Major aorta pulmoner kollateral arter

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INTRODUCTION

Pulmonary atresia (PA) with ventricular septal defect (VSD) and major aorta pulmonary collaterals (MAPCA) is a complex lesion in which considerable morphologic variability exists regarding the sources of pulmonary blood flow. The clinical presentation of these patients is variable, depending on the magnitude of the pulmonary blood flow, which allows some patients to maintain an excellent functional status for many years¹. Treatment goals for patients who have tetralogy of Fallot with pulmonary arterial hypoplasia and major aorto-pulmonary collateral arteries (MAPCAs) are to establish a central source of pulmonary blood flow which is sufficient to allow closure of the VSD with low and stable right ventricular pressure²⁻⁵. The surgical management of this complex anomaly represents a major challenge and should be individualized according to arborization of pulmonary vasculature and amount of pulmonary blood flow⁶. Two main basic concepts have been used in its management. Primary corrections for infants with congenital complex heart diseases have been recently recommended². Some patients, however, require a palliative systemic to pulmonary shunt operation.

Palliative procedures were designed to improve

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patient symptomatology; most of these were systemic to pulmonary artery shunts¹. The staged repair can be successfully used to treat patients with pulmonary atresia with ventricular septal defect and major aorto-pulmonary collateral arteries. This method yields a relatively low mortality with good functional results⁷. The Blalock-Taussig shunt has been performed with low risk and is associated with excellent pulmonary artery growth.

CASE

The patient was 8 year old girl. She admitted to the hospital with mild cyanosis and palpitation. On examination she had easily palpable pulses, there was a grade 3/6 ejection systolic murmur at the upper left sternal edge, mild cyanosis and clubbing. Cardiac and selective ascending aortic catheterization showed multiple abdominal and thoracic MAPCA's, hypoplastic pulmonary artery and confluent pulmonary flow was filling with MAPCA's. Prior to the surgery, two major aorto-pulmonary collateral arteries (abdominal and thoracic) were embolized using steel coils at the same procedure (Figure 1 and 2). The collateral artery from right internal mammarian artery (RIMA) was not embolized due to blood supply for pulmonary arterial circulation until palliative procedure was done (Figure 3).

There was a single large ventricular septal defect in the subaortic position and severe infundibular narrowing of the small pulmonary valve and hypertrophied

Figure 1: Abdominal MAPCA which was occluded with coil embolization

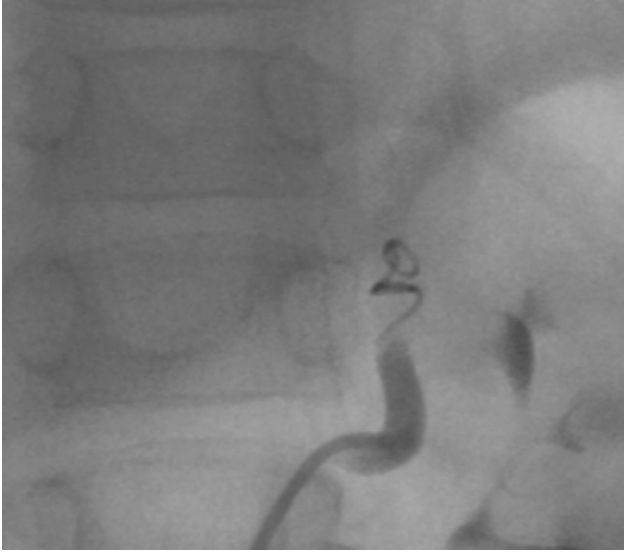
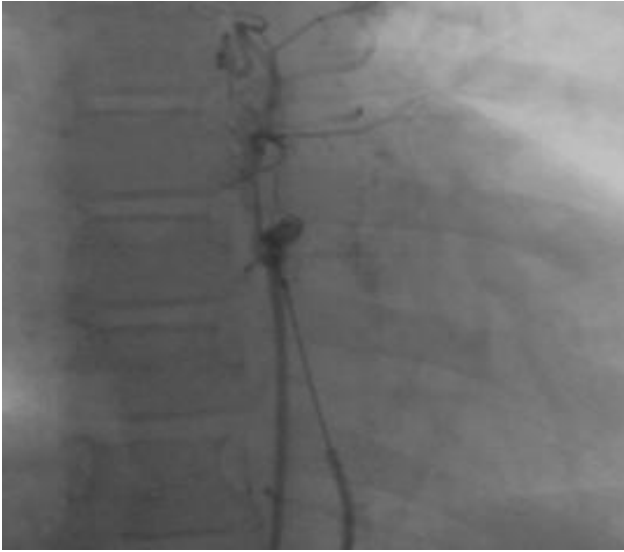
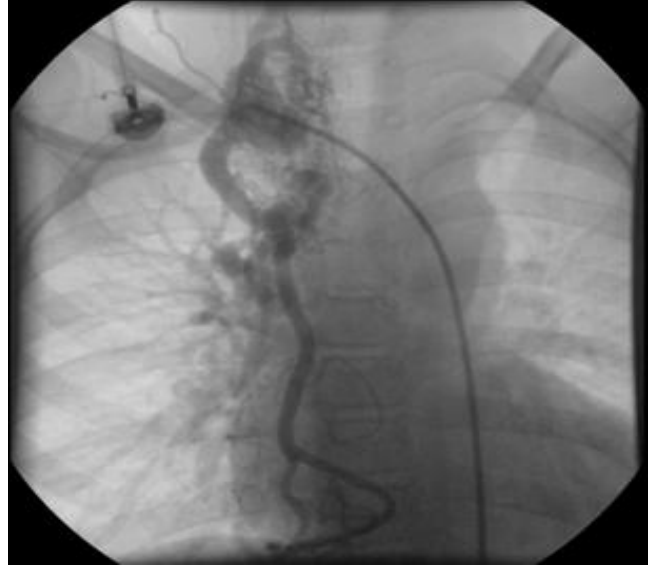


Figure 2: Thoracic MAPCA was occluded with coil embolization



right ventricle. Coronary arteries were thought to be normal. Her chest radiograph showed levocardia with an uptilted apex and decreased pulmonary vascular markings and right-sided aortic arch. Cross-sectional echocardiography demonstrated situs solitus with concordant connections. There was severe tetralogy of Fallot and severe pulmonary arterial tree hypoplasia. The systemic and pulmonary venous drainage were normal. McGoon index was calculated as 1.1. Palliative surgical procedure was undertaken during the same admission. After general anesthesia the right hemithorax was entered through the fourth

Figure 3: The angiography showing systemic to pulmonary arterial circulation from RIMA



interspace. The huge collateral circulation from RIMA was seen. After systemic to pulmonary arterial shunt (modified BT shunt) was performed with 5mm PTFE (Gore-Tex; W. L. Gore & Associates, Inc, Flagstaff, Ariz.) graft and the collateral artery was transected and sutured in a running fashion.

One very unusual aspect of this patient's anatomy was the origin of the collateral circulation from RIMA.

DISCUSSION

The adequate blood flow is an important factor in the growth and development of the pulmonary arteries. The true central pulmonary arteries range from a size approaching normal to complete absence. The blood flow is dependent upon the severity of right ventricular outflow tract obstruction or pulmonary arterial hypoplasia and the presence of additional blood supply like PDA, systemic to pulmonary shunts or MAPCAs⁸. In patients with severe cyanosis, development of minor systemic to pulmonary collateral vessels is commonly seen. Major aorto-pulmonary collaterals, probably derived embryologically from the splanchnic vascular plexus¹⁰, are also highly variable in their size, number, course, origin, arborization. Heterogeneity of anatomical patterns of pulmonary blood supply has always provided ground for controversies concerning the optimal surgical strategy for pulmonary atresia with ventricular septal defect and multiple aorto-pulmonary collateral arteries (PA, VSD, MAPCAs)⁹. In this case there were several different blood source for pulmonary circulation.

According to our knowledge there is no such a blood source for tetralogy of Fallot with pulmonary hypoplasia from RIMA in English literature.

The integrity of the pulmonary vascular bed is a major issue in the management of pulmonary atresia with MAPCAs. MAPCA is not a safe blood source for pulmonary circulation. It is essential to create a safe blood supply for pulmonary arterial bed with a shunt. Most staged approaches use circumferential nonviable conduits in the central and peripheral pulmonary circulation. The anatomy of the pulmonary circulation in patients with tetralogy of Fallot and major aortopulmonary collateral arteries is usually less complex than in pulmonary atresia with ventricular septal defect, making the decision for complete repair more hopeful. In this case our major concern was pulmonary vascular bed and palliative procedure was chosen due to severe pulmonary hypoplasia (McGoon index was 1.1). The patient was in a good postoperative condition after surgery. A coil embolization is useful for MAPCAs which located different places.

For tetralogy of Fallot with pulmonary atresia or hypoplastic or absent pulmonary arteries and major aortopulmonary collateral arteries, a staged surgical approach yields low overall mortality and acceptable hemodynamics after complete repair.

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