Cardiology in the Young

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An unusual case of pulmonary atresia with ventricular septal defect and multiple major aortopulmonary collateral arteries: undiagnosed until adulthood

Feyza Sönmez Topcu[1], Baran Simsek[2] and Arda Ozyuksel[2,3]

[1]Department of Radiology, Aydin University, Medical Park Florya Hospital, Istanbul, Turkey; [2]Department of Cardiovascular Surgery, Medicana International Hospital, Istanbul, Turkey and [3]Department of Cardiovascular Surgery, Biruni University, Istanbul, Turkey

Brief Report

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Author for correspondence: Baran Simsek, Department of Cardiovascular Surgery, Medicana International Hospital, Istanbul, Turkey. E-mail: simsekbaran7@gmail.com

Abstract

Pulmonary atresia and ventricular septal defect with major aortopulmonary collateral arteries is an extremely complex, heterogeneous, and rare anomaly. This group of patients may not be able to survive until adulthood without any interventions or treatment. Although surgical management of patients diagnosed in newborn, infant, or early childhood is clear, treatment of patients diagnosed in adulthood still remains a significant problem. The pre-operative clinical status, imaging methods, and operative findings might be helpful for planning the most appropriate management. Herein, we report a unique case of pulmonary atresia and ventricular septal defect with major aortopulmonary collateral arteries who remained asymptomatic until the age of 18 years.

Case report

An 18-year-old female, without prior diagnosis of heart disease presented with the complaint of dyspnea on exertion. Due to the religious and conservative pattern of the patient's family, she did not apply to a hospital even if for a basic physical examination until the patient was symptomatic. Physical examination revealed grade III clubbing with a resting oxygen saturation of 72%. Cardiac auscultation revealed systolic murmur in the precordium. Transthoracic two-dimensional echocardiography revealed a single, large, malaligned non-restrictive ventricular septal defect, pulmonary atresia with hypoplastic and confluent branch pulmonary arteries, multiple major aortopulmonary collateral arteries, and a right aortic arch. Both ventricles were enlarged but preserved their functions. CT pulmonary angiography revealed a hypoplastic main pulmonary artery with bilaterally hypoplastic branch pulmonary arteries with a non-visualised right ventricular outflow tract (Fig 1). There were three major aortopulmonary collateral arteries rising from the descending aorta, one from the right subclavian artery and two from the left subclavian artery, respectively (Fig 2). The left brachiocephalic vein was retroaortic. The aortic arch was right-sided. Diagnostic cardiac catheterisation revealed one major aortopulmonary collateral artery originating from the right subclavian artery which was 7 mm in diameter and in association with the hypoplastic main pulmonary artery, two major aortopulmonary collateral arteries originating from the left subclavian artery which were 3 and 4 mm in diameter and three major aortopulmonary collateral arteries which were 4.5, 3.5, and 3 mm in diameter and originating from the descending aorta, respectively. The largest major aortopulmonary collateral artery was originating from the right subclavian artery and was associated with the hypoplastic main pulmonary artery with a pressure value of 34/16 (25) mmHg. The second largest major aortopulmonary collateral artery was the topmost of the three major aortopulmonary collateral arteries originating from the descending aorta and was predominantly supplying the left lung with a pressure value of 32/15 (24) mmHg. The left ventricular pressure value was 80/0–9 mmHg at the same time. We could not measure the central pulmonary artery pressure. Right pulmonary artery was 7 mm in diameter, whereas the left pulmonary artery was 8 mm.

Under standard cardiac anesthesia, following the median sternotomy, total excision of the thymus was performed. Pericard was opened wide enough to provide exposure of the aorta and the pulmonary arteries. The ascending aorta and the pulmonary arteries were mobilised. 100 units/kg intravenous heparin was administered and after confirming an activated clotting time over 200 seconds, Castaneda clamps were positioned, and a 4-mm expanded polytetrafluoroethylene vascular graft was interposed between the lateral side of the ascending aorta and the left pulmonary artery with 7/0 polypropylene sutures, respectively. Aortic anastomosis was in a side-to-side fashion, where the left pulmonary artery anastomosis was end to side. The most

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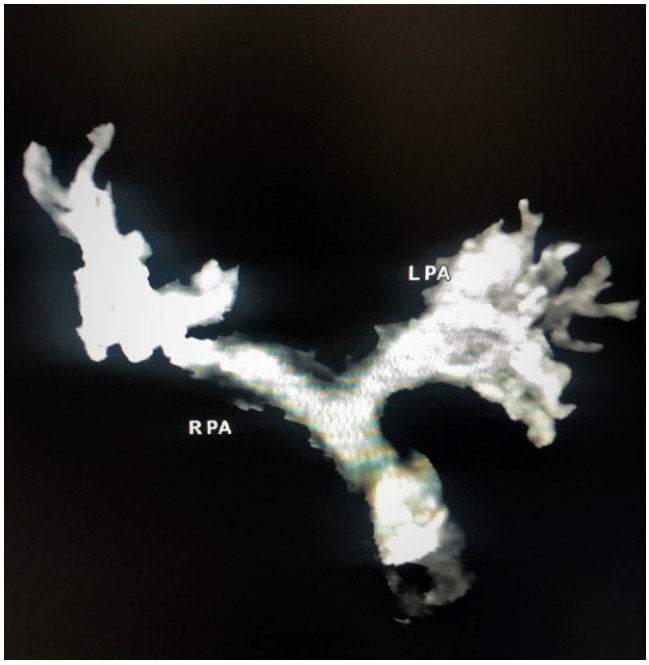

Fig. 1 Three-dimensional CT revealing the hypoplastic right and left pulmonary arteries.

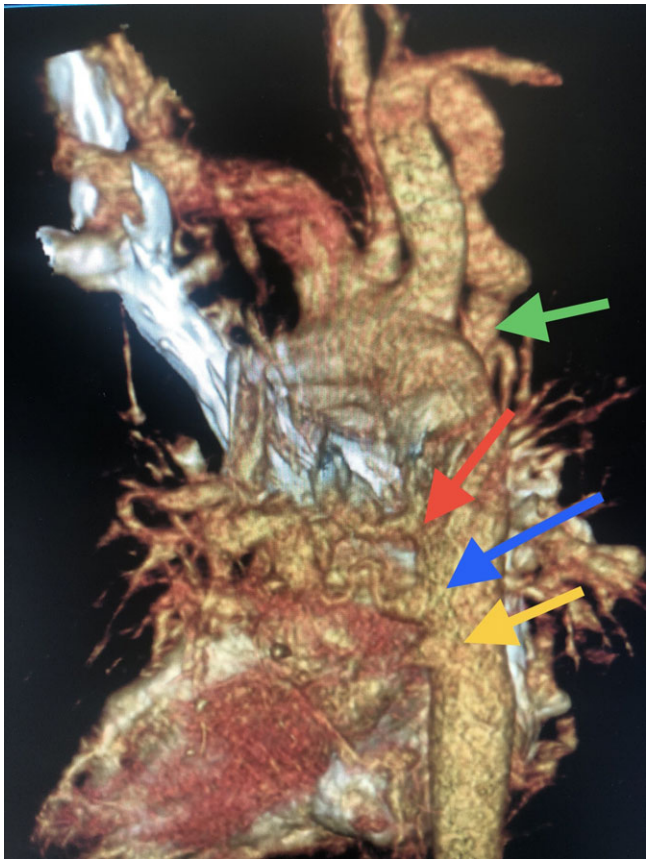

Fig. 2 Three-dimensional volume-rendered image.

proximal part of the graft stump was folded and closed with double row 6/0 polypropylene sutures and secured with two surgical clips. The central aortopulmonary shunt procedure was successfully performed without cardiopulmonary bypass support. After completion of the anastomosis and removal of the Castaneda clamps, systemic arterial oxygen saturation level was 89% under fraction of inspired oxygen of 50%. Heparin infusion with a dosage of 1 mg/kg/hour was initiated during the first 72 hours post-operatively based on the activated clotting time values. Afterwards, total daily heparin dosage was administered intravenously in four divided injections per day. Oral aspirin was administered at 150 mg/day beginning from the first post-operative day. The patient's post-operative course was uneventful and discharged on the sixth post-operative day. A future total correction procedure due to a satisfactory development of the pulmonary vascular bed should be considered; however, the shunt procedure should be a long-term palliation leading to a better quality of life. The future evaluation of the patient's physiology will be decisive to determine whether she is a potentially candidate for a repair.

Discussion

Pulmonary atresia and ventricular septal defect with major aorto-pulmonary collateral arteries occurs in approximately 10/100.000 live births and represents a complicated anatomic and surgical spectrum of CHD.[1] Aortopulmonary collaterals develop due to the absence of antegrade pulmonary blood flow. In the absence of a patent ductus arteriosus, aortopulmonary collaterals will be the sole source of pulmonary blood flow. The collaterals which provide different amounts of blood flow to different pulmonary segments are anatomically variable and prone to stenosis over time. The majority of aortopulmonary collateral arteries originate from the descending aorta but have also been found arising from the aortic arch, the left coronary artery, subclavian artery, internal mammary artery, and the distal thoracic aorta.[2] The importance of early definition of pulmonary blood supply is paramount, establishing which areas of the lung are supplied by major aortopulmonary collateral arteries alone and which have dual supply with the native system. In patients with dual-supply major aortopulmonary collateral arteries and confluent but hypoplastic central pulmonary arteries, our surgical approach is to perform a shunt procedure to promote pulmonary artery growth. Rehabilitation of small native vessels with central shunts should be very effective.

Without intervention, the survival rate of this disease is low, with a 10-year and 20-year survival rates reported at 50 and 10%, respectively.[1,3] Establishing the sources of pulmonary blood flow and creating suitable pulmonary arborisation are principles to manage this anomaly. There is a controversy regarding the optimal treatment of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Different strategies have been performed, such as providing right ventricle to pulmonary artery continuity by single or multistage approaches, with or without intracardiac repair, central aortopulmonary shunting with unifocalisation, and creation of an aortopulmonary window regarding the suitability of the length of aortopulmonary collaterals, existence of stenotic segments and native pulmonary arteries, and the status of distal pulmonary vascular bed.[4–6]

Melbourne and Stanford groups defined two major approaches for the surgical technique.[4,5,7] According to the Melbourne group, unifocalisation is not deemed mandatory due to thrombosed or narrowed unifocalised vessels over 50% at a mean of 3.4 years with a complete repair rate of 73%.[8] In contrast, the Stanford group's algorithmic approach was based on single-stage total correction.[9] When small central pulmonary arteries are present and seem to be distributed to most of both lungs, it is recommended to grow these

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pulmonary arteries by either a central shunt, a Melbourne shunt, or restoration of the right ventricle to pulmonary artery continuity.[10] Patients with small central pulmonary arteries may be better managed by leaving the ventricular septal defect open and taking advantages of shunts or limiting right ventricle to pulmonary artery conduits as long-term palliation leading to a very good long-term quality of life in selected patients.[10] On the other hand, the shunting procedures have been particularly successful in developing even diminutive native pulmonary arteries, and the long-term survival of the patients and their quality of life seemed to depend mainly on the growth of the native vessels by these shunting procedures.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the Helsinki Declaration of 1975, as revised in 2008.

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