Simple transposition of the great arteries with total anomalous pulmonary veins connection (infracardiac form): case report

Transposición simple de las grandes arterias con conexión de venas pulmonares anómalas totales (forma infracardíaca): reporte de caso

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Abstract

Transposition of the great arteries (TGA) with total anomalous pulmonary veins connection (TAPVC) is an extremely rare clinical observation. Up today in medical literature 8 similar cases were found. There are mostly presented the combinations of TGA with supra- and intracardial forms of TAPVC. In two cases was performed anatomic correction of the TGA with no obstructive form of infracardiac and intracardiac TAPVC. The aim of our report is to present the successful anatomic correction in infant with d-TGA and infracardiac form of total anomalous pulmonary veins connection into the portal vein with stenosis of the pulmonary vein collector.

Key words: transposition of the great arteries, total anomalous pulmonary veins connection, arterial switch operation.

Resumen

La transposición de las grandes arterias (TGA) con conexión de venas pulmonares anómalas totales (TAPVC) es una observación clínica extremadamente rara. Hasta hoy en la literatura médica se encontraron 8 casos similares. En su mayoría se presentan las combinaciones de TGA con formas supra e intracardíacas de TAPVC. En dos casos se realizó corrección anatómica de la TGA sin forma obstructiva de TAPVC infracardiaca e intracardiaca. El objetivo de nuestro informe es presentar la corrección anatómica exitosa en lactantes con d-TGA y la forma infracardíaca de la conexión total de las venas pulmonares anómalas en la vena porta con estenosis del colector de la vena pulmonar.

Palabras clave: transposición de las grandes arterias, conexión de venas pulmonares anómalas totales, operación de cambio arterial.

Introduction

Combination of transposition of the great arteries (TGA) with the total anomalous pulmonary venous connection (TAPVC) is an extremely rare clinical observation. Literature sources present rare cases of the combination of TGA with supra- and intracardiac forms of TAPVC[1-5]. Only several publications describe the combination of TGA with an infracardiac form of TAPVC. The first report about successful single-stage anatomical correction of TGA with no obstruction form of the infracardiac TAPVC was published

by Lopes L.M. et al[6]. We report a case of stage-by-stage treatment of infant with TGA and obstructive form of infracardiac total anomalous pulmonary venous connection.

The newborn boy with a body mass of 3.5 kg was brought to the neonatal emergency surgery department with simple TGA. When the baby was admitted, his state was considered as extremely severe. The tachypnea was up to 70 breaths per minute. The pulse filling was satisfactory on all the limbs and the arterial pressure had no systolic gradient. The oxygen saturation of the capillary blood was 78-83%. The patient was infused PGE-1 preparation. Echocardiography diagnosis was O-TGA, restrictive patent foramen ovale with a right-to-left blood flow, patent ductus arteriosus 4.3 mm with a predominantly right-to-left blood flow. The indexed end-diastolic volume of the left ventricle was 15 ml/m^2; indexed LV myocardial mass was 27 g/m^2; the diameter of the mitral valve was 9 mm. TGA was associate with total anomalous pulmonary venous connection to the portal vein with stenosis of the collector of the pulmonary veins (the systolic pressure gradient was 20 mm Hg). In order to examine the anatomy of the pulmonary veins, the multispiral computed tomography with contrast agent was performed. All the pulmonary veins had gathered into a common collector with a diameter of 6.0 mm and had joined the portal vein. The collector narrowed to 2.5 mm at the place of the connection (**image 1**). Due to the increase the pulmonary-cardiac failure and the reduction oxygen saturation in arterial blood, the newborn was subjected to artificial ventilation of the lungs and the Rashkind procedure was performed. The size of the atrial septal defect has become 6 mm.

Image 1

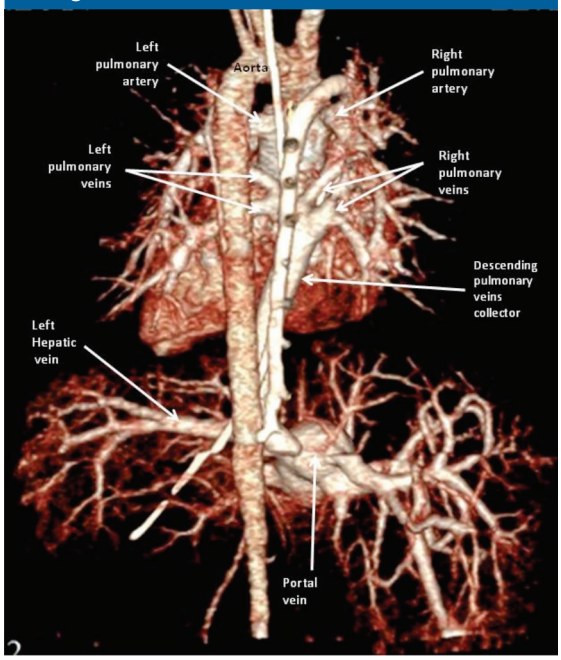

Image 1. A) Multislice computer tomography with contrast agent. 3D reconstruction. All the pulmonary veins had gathered into a common collector and had joined the portal vein. The collector narrowed to 2.5 mm at the place of the connection.

The volume hypoplasia of the left ventricle and the risk of a large-scale surgical injury in the case of the single-stage radical correction made us take the decision about a two-stage treatment of the anomalous. At the age of 15 days old, the newborn underwent an operation in order to "move" the collector of the pulmonary veins to the left atrium, narrow the pulmonary artery and create a systemic to pulmonary anastomosis. An anastomosis was made between the left atrium and the collector of the pulmonary veins under the cardiopulmonary bypass and hypothermia. The distal end of the collector was occluded by a titanium clip. Under the parallel cardiopulmonary bypass, a systemic-to-pulmonary shunt was made with a vessel prosthesis from PTFE (diameter 3.5 mm) between the unnamed artery and the right pulmonary artery. The pulmonary artery was narrowed with a lavsan cuff.

The child was admitted to the hospital again at the age of 9 months. According to the results of the echocardiography and computed tomography angiography, the anastomosis of the collector and the left atrium were of normal size, the blood flow was laminar. An arterial switch operation was performed. The neopulmonary artery root was made from the autopericardium. The infant was extubated on the 2nd day after the operation and was discharged home on the 10th postoperative day.

Discussion

Total anomalous pulmonary veins connection is a rare congenital heart defect. The frequency of TAPVC occurrence among all Congenital Heart Defects is up to 1%[7]. TAPVC is often found as an isolated heart disease, but can be the part of a complex congenital heart disease or heterotaxy syndrome with asplenia. But the combination of TGA and TAPVC is extremely rare. The hemodynamics of simple TGA consists in parallel systemic and pulmonary circulation, and without large fetal shunts, such as patent foramen ovale or patent ducus arteriosus, severe cyanosis and the critical condition of the newborn develops rapidly after birth. However, the combination of TGA with TAPVC and medium size of the patent foramen ovale is a unique natural defect compensation, in which the ratio of pulmonary blood flow to the systemic flow (Qp/Qs) can reaches normal values[8].

Most authors offer as a surgical intervention in a surgical approach to an atrial plane with modified methods of Senning or Mustard to treatment similar combination of congenital heart defects[1-5]. In 1984[1] M. Verberro-Marcial et al. published a report on the case of successful hemodynamic correction of TGA with an intact ventricular septum in combination with the supra-cardiac form of TAPVC for an 8-month-old child weighing 9 kg. The modified Senning procedure was chosen to correct the transposition of large arteries. Thies WR et al., 1990[9],

described a case of a 4.5-month-old baby weighing 4.5 kg, in which a TGA with an infracardiac form of TAPVC, a blockage of the collector and a left atrial membrane was hemodynamically corrected. The operation "Mustard" was performed, the left atrium membrane was cut out, and the anastomosis of the pulmonary vein collector with the left atrium was made.

The authors were justified the choice of hemodynamic correction of concomitant defect by the small size of the LV and the inability to perform ASO. Indeed, the combination of these rare congenital heart defects can lead to a rapid decrease the LV volume during the first two weeks after birth. This period is an optimal time for anatomical surgery in patients with TGA. The factors affecting the regression of the LV mass with simple TMA include 1) early closure of the patent ductus arteriosus. The mechanism is a decrease in the LV afterload, and 2) a large atrial septal defect (natural or after balloon atrial septostomy) decrease the LV preload. Some authors recommend to avoid to perform the ASO in patients with TGA add LV indexed myocardial mass less than 35 g/m^2 [10]. That was done in our case. Firstly, pulmonary veins were connected to the left atrium, and then was performed pulmonary artery banding and the systemic-to-pulmonary artery shunt. And ASO was successfully performed in patient at age of 9 months. Over the past 5 years, we have found only the one report of the successful anatomical correction in patient with TGA and TAPVC to the coronary sinus[2].

Patients with combination of TGA and TAPVC have higher values of the oxygen saturation in the, particularly in the presence of a restrictive patent foramen ovale and/or in the absence of patent ductus arteriosus. Our patient had a restrictive patent foramen ovale, right-to-left shunt on patent ductus arteriosus and the oxygen saturation of the capillary blood was reached 83%. Echocardiography is the primary method of the instrumental diagnostics in pediatric cardiology. So, the clinical and ultrasound data should be compared for full and timely diagnosis.

Therefore, our case report has a clinical interest due to the rarity of the congenital heart defect combination (TGA and obstructive form of infracardiac TAPVC) and the choice of surgical treatment strategy. The description of our case proves once again that we should strive to perform the anatomical correction of the TGA even in combination with TAPVC.

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