Favorable Postoperative Evolution after Late Surgical Repair of Truncus Arteriosus Type 1: A Case Report

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ABSTRACT

Truncus arteriosus communis (TA) is a rare cyanotic congenital heart defect, in which the aorta and the pulmonary artery have not been separated during the normal development of the fetal heart, so a single truncal artery is arising from the base of the heart. Most patients with TA present within the first weeks of life with heart failure. This anomaly is an important cause of pulmonary arterial hypertension. Corrective surgery is indicated in the first 3 months of life, to avoid the development of severe pulmonary arterial hypertension. We report the case of a 12-month-old male infant diagnosed by echocardiography with truncus arteriosus type 1 in whom, based on hemodynamic data, surgical treatment could be performed at the age of 1 year.

Keywords: truncus arteriosus, pulmonary arterial hypertension, children

INTRODUCTION

Truncus arteriosus (TA) is a rare cyanotic congenital cardiac malformation characterized by a single common artery, which emerges from the heart through a single semilunar truncal valve and delivers blood to the systemic, pulmonary, and coronary circulation. Pulmonary arteries emerge from the common arterial trunk, after the origin of the coronary arteries and proximal to the first brachiocephalic branch of the aortic arch.¹⁻³ Characteristically, there is a misalignment over a large ventricular septal defect. This anomaly is often associated with various disorders such as truncal valve stenosis or regurgitation, coronary artery anomalies, atrial septal defect, aberrant subclavian arteries, persistent superior vena cava and a patent ductus arteriosus.

There are several clinical classifications that describe the various forms of TA; the most commonly used are the Collett and Edward classification and the Van Praagh classification.²

Truncus arteriosus is a life-threatening condition, as its natural history bears a mortality rate of 80% in the first year of life and especially during infancy.² The

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FIGURE 1. Echocardiography (2D and color Doppler) – subcostal view, showing a single overriding great vessel arising from the heart and the truncus of pulmonary artery emerging from the common arterial trunk. MPA – main pulmonary artery, LPA – left pulmonary artery, RPA – right pulmonary artery, AO – aorta

pathophysiology and the clinical manifestations of TA are particularly dependent on the volume of pulmonary blood flow, assessed by the pulmonary vascular resistance (PVR) and the degree of truncal valve regurgitation. There is mixing of systemic and pulmonary blood at the intracardiac level, resulting in mild or moderate cyanosis. In the first weeks of life, PVR decreases to normal values, leading to pulmonary fluid overload and heart failure. The severity of heart failure is increased in cases with significant truncal valve regurgitation. Corrective cardiac surgery is recommended in the first 3 months of life. In surgically uncorrected cases, pulmonary vascular obstructive disease may develop after the age of 6 months.

CASE REPORT

We report the case of a 12-month-old male infant who was referred to our department with a history of respiratory distress, difficulty in breathing, tachypnea, poor feeding, and failure to thrive. On admission, the clinical examination revealed mild cyanosis, peripheral oxygen saturation was 94%, respiratory rate was 70 respirations per minute, heart rate was 150 beats/minute, blood pressure was 90/48 mmHg, and there was a loud and single second heart sound on auscultation and a harsh systolic murmur (3/6) at the lower left sternal border. The liver was enlarged, approximately 2 cm below the right costal margin. His weight was 6.5 kg (80% of the expected weight). The hemoglobin concentration was 12.2 g/dL, and the hematocrit value was 34%.

The electrocardiogram showed peaked P wave and bi-ventricular hypertrophy, and the chest X-ray showed cardiomegaly with prominent right cardiac cavities and increased pulmonary vascular markings.

The echocardiography demonstrated the typical findings of TA: a single arterial stem giving rise to the pulmonary trunk with normal-sized pulmonary arteries arising from the main pulmonary artery segment (Figure 1). A large ventricular septal defect with bidirectional shunt was present. The echocardiographic examination revealed also a quadricuspid truncal valve with mild regurgitation, a normal origin of the coronary arteries, and a normal aortic arch.

The computed tomography (CT) examination confirmed the diagnosis of TA type I (Figure 2A, 2B).

Considering the pathophysiology of this complex congenital heart disease and that the diagnosis was made at the age of 12 months, a hemodynamic investigation was indicated to evaluate the pulmonary pressure and the PVR. The cardiac catheterization revealed quasi-systemic pressures in the pulmonary circulation with a mean pulmonary artery pressure of 47 mmHg, with reactive PVR (3.82 UW/m^2 in atmospheric air and 2.25 W/m^2 after administration of nitric oxide). These findings indicated the need for surgical correction.

A standard method of TA repair was performed with the termination of the ventricular septum defect, separation of the pulmonary arteries from the primitive truncus, and restoration of the right ventricle and pulmonary artery continuity using a 15 mm Contegra conduit. The

FIGURE 2. Computed tomography (**A** – sagittal image; **B** – 3 D reconstruction) showing a truncus arteriosus communis type I image with a single arterial trunk (TA) giving rise to pulmonary trunk which in turn gave rise to right (RPA) and left pulmonary artery (LPA). AO – aorta

surgery was uneventful. Postoperatively, the patient presented symptoms that indicated a low cardiac output due to biventricular dysfunction, requiring the administration of inotrope agents, and pulmonary arterial hypertension (PAH) crises, requiring pulmonary vasodilator therapy consisting of nitric oxide and sildenafil, a phosphodiesterase type 5 inhibitor.

The patient was discharged 20 days after cardiac surgery in optimal condition. The echocardiogram at the time of hospital discharge showed the pulmonary bioprosthesis with a gradient of 12 mmHg and mean regurgitation, mild aortic regurgitation, a ventricular septal defect patch with a small residual left-to-right shunt, and mild tricuspid regurgitation with a gradient of 40 mmHg, with recovery of biventricular function.

The echocardiographic examination performed at 1 year after the corrective cardiac surgery showed good function and no signs of residual pulmonary arterial hypertension.

DISCUSSIONS

TA is an uncommon lesion, with a prevalence of 0.056 to 0.03 in every 1000 births, described as a single arterial stem emerging from the heart, which in turn gives rise to the pulmonary trunk in various ways.^{4–8} TA belongs to the group of ectomesenchymal tissue migration anomalies caused by the aberrant migration of neural crest cells through the branchial arch vessels during cardiogenesis.^{9,10}

Chromosomal abnormalities are detectable in 8.7% of cases of TA. Di George syndrome and 22q11 chromosome microdeletion were reported in more than 30–50% of cases.^{4,11–13} In our report, there was no chromosomal disorder present.

Several cases of prenatal diagnosis have been reported, but in most cases the diagnosis is made during the neonatal period,^{14–16} when it is important to differentiate TA from other cardiac disorders that cause early heart failure and

neonatal sepsis. Failure to thrive, respiratory infections, dyspnea, cyanosis, and clinical signs of congestive cardiac failure are the common features. The electrocardiogram usually shows right axis deviation and right ventricular preponderance, and the chest radiography typically shows cardiac enlargement with an increase in the pulmonary vascular markings.

Diagnosis is achieved by echocardiography, which can identify the type of truncus, the morphology and functionality of the truncal valve, and the physiologic consequences of the cardiac disease.⁸

Due to the advantages of echocardiography, cardiac catheterization with angiography is indicated when pulmonary vascular disease is suspected and to evaluate associated lesions.

The hemodynamic consequences of a common TA may predispose to the development of pulmonary arterial hypertension; for this reason, the corrective operation is indicated before the age of 3 months in order to avoid the development of severe pulmonary vascular obstructive disease.^{3,17} Niwa *et al.* reported a one-year survival rate of over 80% in patients who received surgical correction during the neonatal period.^{18–23} A delay in the surgical treatment is associated with the risk of postoperative PAH crisis and cardiac failure.^{22,24,25} Porter *et al.*, in a case series of uncorrected patients, reported a mean period of survival of 5 weeks and a survival rate of only 15% at the age of 1 year.²⁶ Marcelletti *et al.* concluded that all patients who survive beyond the first year of life, will develop pulmonary vascular obstructive disease.^{18,27}

During the postoperative assessment, a careful follow-up for pulmonary hypertension and truncal conduit patency is needed.

The reported case is one of the few cases diagnosed with TA reported in the literature, in which surgical correction could be performed at the age of 1 year because hemodynamic data did not document the presence of pulmonary vascular obstructive disease. Pulmonary arterial hypertension could seriously complicate the postoperative course of pediatric patients. Although the PAH crises complicated the immediate postoperative evolution, the subsequent evolution of the patient was favorable, and the echocardiography performed at 1 year after surgery indicated no significant residual lesions.

ETHICAL CONSIDERATIONS

The manuscript is coherent with the ideologies stated within the Declaration of Helsinki. The patient's legal guardian signed an informed agreement and agreed to the publication of his data. The study has been approved by the

ethics committee of the University of Medicine and Pharmacy of Tîrgu Mureş, Romania.

CONCLUSIONS

Truncus arteriosus is a complex cardiac disease, in which primary surgical repair should be performed during the neonatal period in order to avoid the development of severe PAH. The diagnosis is generally confirmed by echocardiography. The management should be individualized according to the age of the patient, the anatomical type of the TA, the associated lesions, and the hemodynamic consequences.

CONFLICT OF INTEREST

None declared.

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