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Truncus arteriosus with meandering pulmonary arteries

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The authors stated that the written consent was obtained from the parents of the patient presented with images in the study.

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Abstract

Truncus arteriosus is a rare, cyanotic, and congenital heart defect occurring due to failure in the differentiation of the aorta and the pulmonary artery during fetal development. The disease is categorized into four sub-categories in the Van Praagh and Collett & Edwards classification systems according to the origin of the pulmonary arteries. Surgical correction of the pulmonary arteries and repair of the ventricular septal defect is the preferred treatment strategy of choice; this intervention is required early in life. Here, we report a four-month-old baby with truncus arteriosus consisting of atypical pulmonary anatomy undefined by either the Van Praagh or the Collett & Edwards classification systems who underwent successful corrective surgery.

Keywords: truncus arteriosus, pulmonary artery, classification, surgical treatment

Introduction

Truncus arteriosus (TA) is a rare congenital cardiac defect encompassing 1.4%-2.8% of congenital heart diseases [1]. TA consists of a common arterial trunk superior to the ventricular septal defect. The solitary truncal artery is the only systemic, pulmonary, and coronary circulation supplier, and the aorta of such patients is wider than normal . The aortic valve and arch anomalies are common in patients with TA [2]. Nevertheless, the cardiac defects most related to TA include tricuspid valve dysplasia or dysfunction, ventricular septal defects (VSD), aortic arch abnormalities, and coronary artery malformations [3].

Patients with TA frequently have a clinical presentation of dyspnea, recurrent lung infections, and growth retardation. Symptoms may range from dyspnea to co-incidental findings on radiographs. Imaging methods include echocardiography, angiography, computed tomography, or magnetic resonance for screening and diagnosis. After diagnosing TA, the pathology is classified according to Collett & Edwards or the Van Praagh classification systems. These classification methods are based on the anatomical structure of the left pulmonary artery, right pulmonary artery, and aorta. The surgical treatment strategy is also planned according to the type of TA. Our article focuses on the diagnosis and surgical management of a 4-month-old male patient with findings of an atypical TA with meandering pulmonary arteries.

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Case presentation

Here, a 4-month-old male patient without an antenatal or postnatal diagnosis was referred to our institution with increasing dyspnea, easy fatigability, poor feeding, and growth retardation. His parents reported recurrent pulmonary infections and bouts of cough . The patient weighed 3800 g (birth weight: 3250 g) and was 60 cm tall. The body surface area was calculated as 1.24 m^2. On physical examination, pulmonary rales were present, the heart was hyperdynamic with a 2-3/6 systolic murmur at the mesocardiac focus, and the patient had slight hepatomegaly. The patient's blood pressure was 81/43 mmHg. Electrocardiography showed tachycardia (180 beats/min), normal sinus rhythm with regular intervals, and a normal QRS axis. The cardiothoracic index increased with significant bilateral hilar opacities on plain chest X-ray (Figure 1). Following physical examination, there was a high suspicion of congenital cardiac anomaly, which directed us to perform echocardiography. Echocardiography showed truncus arteriosus with a peri-membranous outlet VSD, right-sided aortic arch, a patent foramen ovale with a right to left shunt, and pulmonary hypertension. The aortic valve was tricuspid and exhibited trivial regurgitation. Both pulmonary arteries were present; however, neither their origin nor course could be identified. Therefore, the classification of pathology could not be determined. Anti-congestive therapy with digoxin, captopril, and furosemide was immediately initiated. Diagnostic cardiac catheterization suggested TA. Although there were two well-developed pulmonary arteries, their origins could not be identified (Figure 2). The patient was scheduled for corrective surgery.

After describing the pathology, the patient's parents were informed about the risks and benefits of treatment alternatives. Consent for the procedure and use of the materials for academic purposes was acquired. Following a median sternotomy, the pericardium was excised and harvested. An anatomical examination exhibited a truncal pulmonary artery originating on the anterior aspect of the ascending aorta with a short course dividing into the right and left branches. The right pulmonary branch coursed left and followed a path inferior to the ascending aorta before reaching the right lung. The left branching pulmonary artery curled to the left inferior to the right pulmonary artery before going to the left lung (Figure 3). Pulmonary arteries were prepared and looped. Extracorporeal circulation was initiated following aortic and bicaval cannulation. Cardiac arrest was achieved at 32°C with an antegrade hypothermic Del Nido cardioplegia infusion. The truncal pulmonary arteries were snared and resected from the ascending aorta, and the ascending aorta was repaired with an autologous glutaraldehyde-treated pericardial patch. A right ventricular infundibular incision was made, and the ventricular septal defect was patched with an autologous glutaraldehyde-treated pericardial patch.

The right ventricular outflow tract was reconstructed with a 12 mm bovine jugular vein graft (Contegra, Medtronic PLC, 710 Medtronic Parkway Minneapolis, MN 55432–5604 USA). The graft was anastomosed to the right ventricle and the branching pulmonary arteries after anatomically correcting the meandering pulmonary arteries. The patient was weaned off cardiopulmonary bypass with a moderate dose of inotropic support (0.75 mcg/kg/min milrinone, 0.1 mcg/kg/min adrenaline, and 0.1 mcg/kg/min noradrenaline), and the operation was finalized. The total cross-clamp and cardiopulmonary bypass periods were 95 min and 108 min, respectively. The patient was hospitalized at the intensive care unit and extubated 16 h after the operation. The patient was transferred to the ward on the fourth day following the procedure. After spending nine days in good condition, the patient was discharged from the hospital and was followed actively. The patient showed normal myocardial function with considerable weight gain and growth.

Figure 1: The cardiothoracic index increased with significant bilateral hilar opacities on plain chest X-ray.

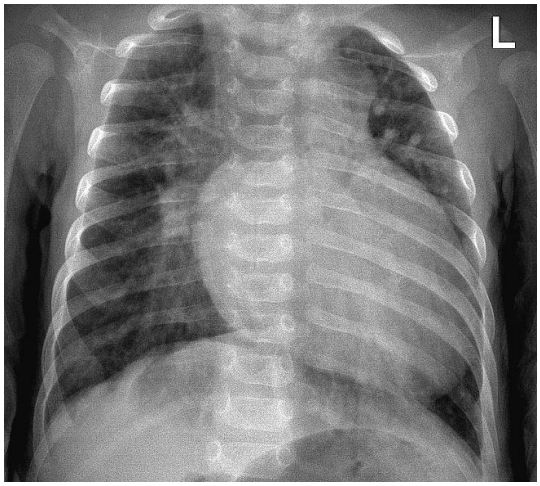


Figure 2: Diagnostic cardiac catheterization indicated two well-developed pulmonary arteries; their origins could not be identified.

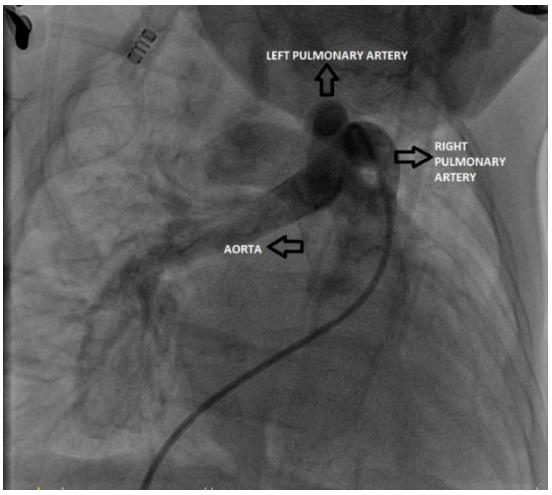
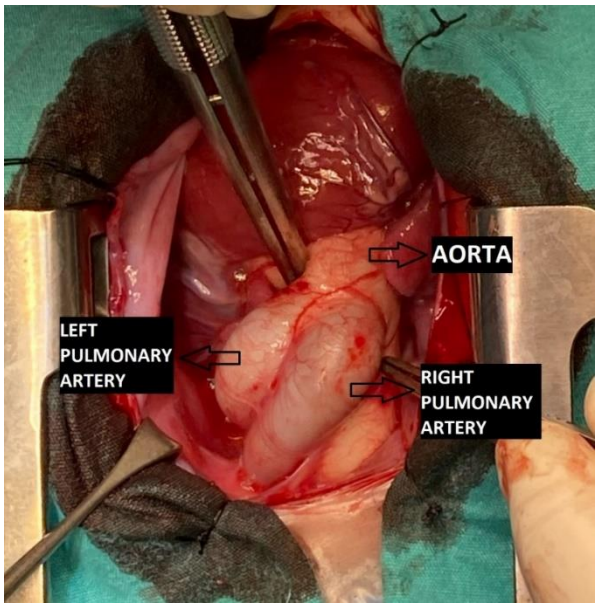


Figure 3: Perioperative view of the main pulmonary artery originating from the anterior surface of the ascending aorta with a concise course that then divides into right and left branches.



Discussion

Truncus arteriosus is a congenital cyanotic heart defect in the pediatric population. It arises from a single truncal valve extending as a single truncal vessel before separating into the aorta and pulmonary arteries. Truncus arteriosus constitutes 4% of critical cases among congenital heart defects. When screening for TA, several anatomical structures are evaluated. These structures include the anatomical orientation and the functionality of the tricuspid valve, the origin of the pulmonary arteries, anomalies of the aortic arch, and the location of the ventricular septal defect [4]. The original and more popular classification of TA was devised by Collett & Edwards in 1949 and divides this anomaly into four types [5]:

- Type I: the truncal pulmonary artery, which splits into right and left pulmonary arteries after emerging from the truncal root (commonly seen in 80% of patients).
- Type II: left and right pulmonary arteries arising separately from the posterior part of the truncus.
- Type III: pulmonary arteries arising from the lateral aspect of the truncus with separate origins.
- Type IV: neither pulmonary arterial branch emerges from the common trunk (pseudo truncus). The pulmonary arteries arise from the descending aorta.

In 1965, Van Praagh modified the classification system to include four primary types [6].

- Type A1: identical to type I proposed by Collett & Edwards.
- Type A2: a separation in the origin of branching pulmonary arteries from the left and right lateral portions of the common trunk.
- Type A3: the branching pulmonary artery (commonly the right) originates from the pulmonary trunk. The branching of the opposite lung is supplied either by collaterals or a pulmonary artery arising from the aortic arch.
- Type A4: interrupted aortic arch coexists with the truncus.

The Van Praagh classification has recently been preferred [8] instead of Collett & Edwards's [7] perhaps because of the inclusion of pulmonary architecture since patients with an underdeveloped aortic arch (interrupted or hypoplastic) with a broad patent ductus arteriosus attached to the descending aorta (15% of the patients with truncus arteriosus) are recognized within the Van Praagh classification [8].

Our patient had a TA consisting of left- and right-branching pulmonary arteries with a conjoined origin separated after emerging from the anterior aspect of the truncal root proximally. This finding is not present in the Collett & Edwards classification or the Van Praagh classification, thus defining the abovementioned architecture as an atypical truncus arteriosus. The atypical formation with such an architecture can be attributed to the embryological development of the heart. The conotruncal septum separates the truncal root into a developed main pulmonary artery and an ascending aorta when the fifth week of gestation concludes. Defects that occur during the formation of the conotruncal septum result in a wide variety of conotruncal abnormalities including TA with a single truncal valve [9].

Neural crest cells and derangements in neural tube development have also been indicated as contributing factors in the development of TA and other conotruncal malformations [10,11]. Apart from developmental abnormalities, environmental and genetic factors also contribute to conotruncal malformations including truncus arteriosus. These risk factors include gestational cigarette smoking [12], advancing maternal age [13], and DiGeorge Syndrome (deletion of 22q11.2) [14]. A genetic screening test with a detailed investigation of the risk factors that the patient or the parents have been exposed to can be performed to further understand the underlying factors that play a role in the formation of atypical cases of truncus arteriosus.

Conclusions

While familial predisposition and risk factors have a significant role in the etiology, the abnormal meandering course and curling of the branching pulmonary arteries could be attributed to a long-standing elevated pulmonary arterial pressure. The architecture of the pulmonary vasculature did not affect the corrective procedure. However, the left and right pulmonary arteries required anatomical alignment because of their unusual presentation in our case. Investigations into similar truncus arteriosus cases can help further explain the underlying etiology of meandering pulmonary arteries.

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