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Echocardiography

A Rare Variant of Type I Truncus Arteriosus: Truncus Arteriosus with Anterior Origin of a Main Pulmonary Artery

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Truncus arteriosus (TA) is an uncommon congenital cardiovascular anomaly that is characterized by a single arterial trunk arising from the normally formed ventricles by means of a single semilunar valve. The anomaly is thought to result from incomplete or failed septation of the embryonic truncus arteriosus. Truncus arteriosus with anterior origin of a main pulmonary artery is a very rare condition. In this report we present a newborn who has a truncus arteriosus with anterior origin of a main pulmonary artery.

Case Report:

A 2250 g infant was born at 38 weeks of gestation and referred to our hospital due to respiratory distress on the first day of life. On admission, systemic oxygen saturation was 90%, respiratory rate was 85 breaths/min., heart rate was 160 beats/min. There was 2/6 systolic ejection murmur present at the left upper sternal border, a systolic ejection click was also noted at the upper left sternal border. ECG showed a right-axis deviation and right ventricular hypertrophy. The chest radiograph demonstrated mild cardiomegaly with pulmonary congestion. Echocardiographic examination showed a normal visceroatrial situs solitus, normal pulmonary venous return, atrioventricular concordance, a common arterial trunk, and a 5 mm subarterial ventricular septal defect (Figs. 1 and 2). The pulmonary artery arose from the anterior surface of the common arterial trunk (Figs. 1 and 2, movie clips S1 and S2). A multislice tomography scan confirmed type 1 truncus arteriosus and pulmonary artery originating anterolateral wall of the truncal artery (Fig. 2). The patient died due to sepsis and lower respiratory infection on the fifth day after admission. Genetic testing for 22q11 deletion could not performed and autopsy could not be carried out due to the lack of the patient's parents' informed consent.

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Discussion:

Truncus arteriosus is a rare anomaly defined as a single great artery that originates from the base of the heart and gives rise to the pulmonary, systemic, and coronary circulation. The anomaly is thought to result from incomplete or failed septation of the embryonic truncus arteriosus. It has an incidence of about 5–9 per 100 000 live births.[1–3] There are several classifications for this anomaly. Van Praagh[1] classified truncus arteriosus into types A and B. In type B, there is no association with a ventricular septal defect. Type A is subdivided as follows: type A1: partially separated pulmonary trunk; type A2: two pulmonary arteries arising directly from the truncus arteriosus; type A3: a single pulmonary artery originating from the arterial trunk, along with collaterals originating from the descending aorta; and type A4: significant abnormalities of the aortic arch in association with anomalies of the ductus arteriosus. Anderson et al. have proposed an alternative simplified classification of common arterial trunk.[3] This group classifies truncus arteriosus into only two categories, namely aortic- or pulmonary-dominant types. Neither Van Praagh nor Anderson report anterior origin of the pulmonary trunk. Also, anterior origin of the main pulmonary artery has not been described in several large series of patients with truncus arteriosus.[4–6]

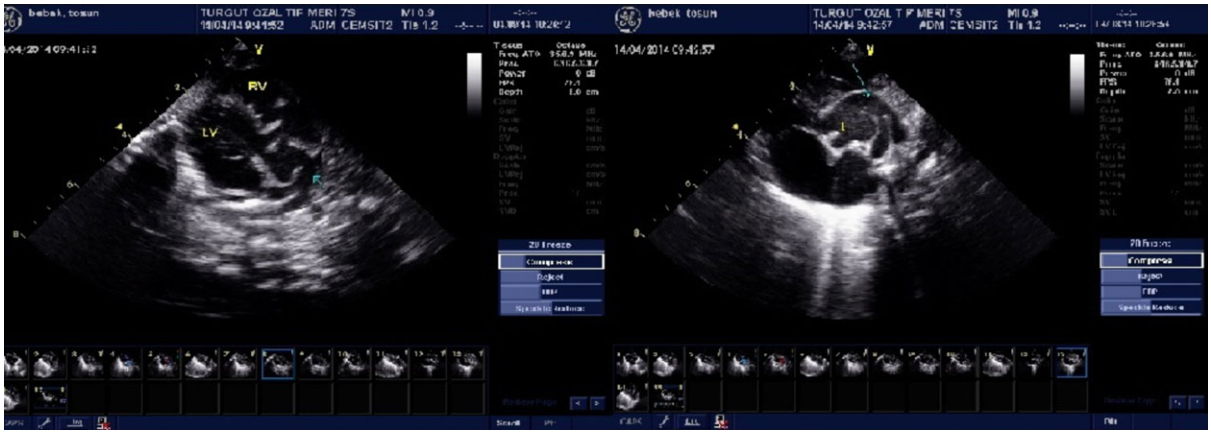

Figure 1. Echocardiographic examination showed a common arterial trunk and a 5 mm subarterial ventricular septal defect. The pulmonary artery arose from the anterior surface of the common arterial trunk.

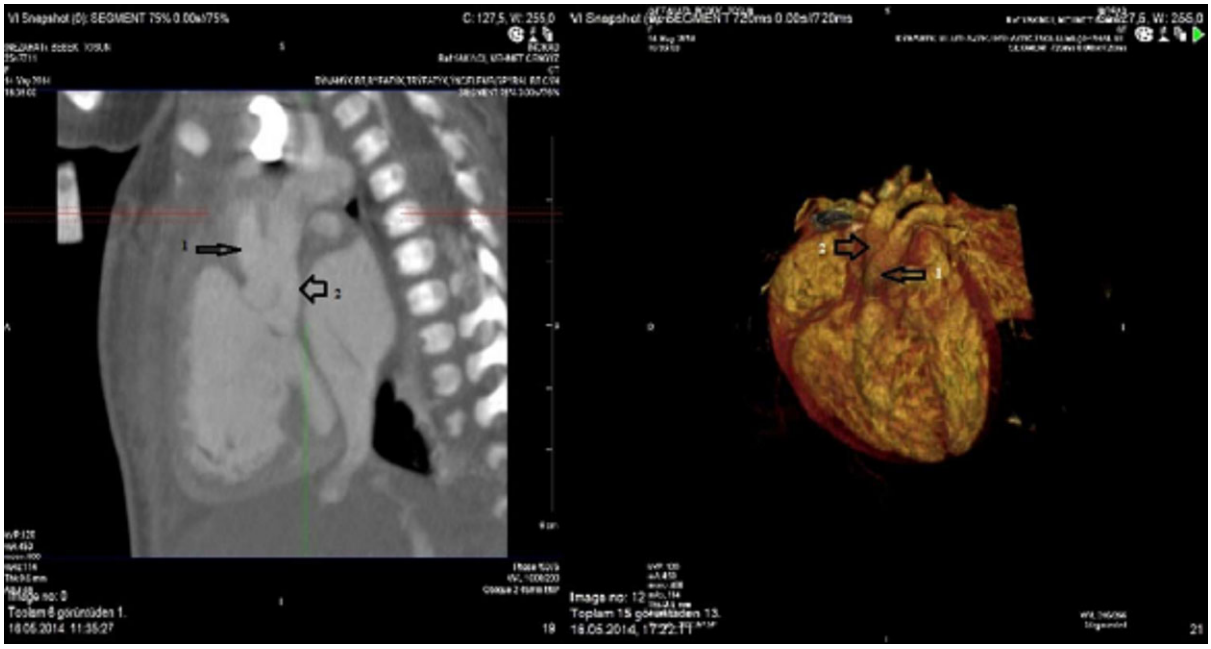

Figure 2. A multislice tomography scan confirmed type 1 truncus arteriosus (2) and pulmonary artery (1) originating anterolateral wall of the truncal artery.

In literature, only two similar cases have been reported as truncus arteriosus with anterior origin of a main pulmonary artery.[7,8] One of them had anterior origin of the main pulmonary artery from the arterial valvar sinus,[7] and the other had truncus arteriosus with anterior origin of a hypoplastic main pulmonary artery.[8] The former patient had undergone complete repair surgery on the seventh day of life and was discharged postoperatively in the second week of life. No further follow-up information about this patient was reported.[7] There is no available clinical information about the second patient, the case report only covers radiological appearance.[8]

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clip S1. Shows pulmonary artery arising from the anterior surface of the common arterial trunk.

Movie clip S2. Shows pulmonary artery arising from the anterior surface of the common arterial trunk. Echocardiographic examination also shows a 5 mm subarterial ventricular septal defect.