The Turkish Journal of Pediatrics 36: 171-174, 1994

A CASE OF COMMON TRUNCUS ARTERIOSUS WITH RARE CARDIAC ABNORMALITIES*

*Hale Ören MD**, Hasan Özkan MD**, Bahattin Ören MD**
*Ali Küpelioğlu MD***, Necla Çevik MD****

SUMMARY: Ören H, Özkan H, Ören B, Küpelioğlu A, Çevik N. (Departments of Pediatrics and Pathology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey). A case of common truncus arteriosus with rare cardiac abnormalities. Turk J Pediatr 1994; 36: 171-174.

Truncus arteriosus communis is a well-known, congenital, cardiac abnormality. In this report, we describe a very rare variant in which the truncus arose from the right ventricle, the atrial and ventricular septa were intact, the left ventricle was hypoplastic and the small right atrium was draining to the truncus arteriosus through a fibrose tunnel-like connection. *Key words: common truncus arteriosus, congenital heart disease, hypoplastic left ventricle.*

Truncus arteriosus communis is defined as a congenital cardiovascular malformation in which one major artery arises from the base of the heart and gives origin to the coronary, pulmonary and systemic arteries[1,2],

In this report, we present a very unusual variant in which the truncus arteriosus arose from the right ventricle, the ventricular and atrial septa were intact, the left ventricle was hypoplastic, and the small left atrium was draining to the truncus arteriosus through a fibrose tunnel-like connection.

Case Report

A three-day-old white male was seen in the emergency clinic for severe cyanosis and respiratory failure. He had a history of mild cyanotic episodes which disappeared spontaneously in a short time. He went into cardiopulmonary arrest immediately after admission to the clinic. He did not respond to cardiopulmonary resuscitation. The electrocardiogram displayed the full pattern of asystoli. No further laboratory investigations, such as chest X-ray, echocardiogram or hemodynamic studies, were performed, as the patient dead on admission.

At necropsy the systemic and pulmonary venous connections were seen to be normal. There was a common arterial trunk, guarded by a three-leaflet common

* From the Departments of Pediatrics and Pathology, Dokuz Eylül University Faculty of Medicine, İzmir.

** Lecturer in Pediatrics, Dokuz Eylül University Faculty of Medicine.

*** Associate Professor of Pathology, Dokuz Eylül University Faculty of Medicine.

**** Professor of Pediatrics, Dokuz Eylül University Faculty of Medicine.

valve which originated solely in the right ventricle. The trunk gave rise to the ascending aorta and, in its posterior aspect, to both pulmonary artery branches and a stenotic arterial duct continuous with the left-sided descending aorta. Two coronary arteries were present, arising from the anterior sinuses. The left atrium was small and the left ventricle was diminutive and poorly developed (Fig. 1). The mitral valve was also rudimentary. The enlarged right atrium was connected to a dominant hypertrophied right ventricle (Fig. 2). The tricuspid valve was dysplastic.

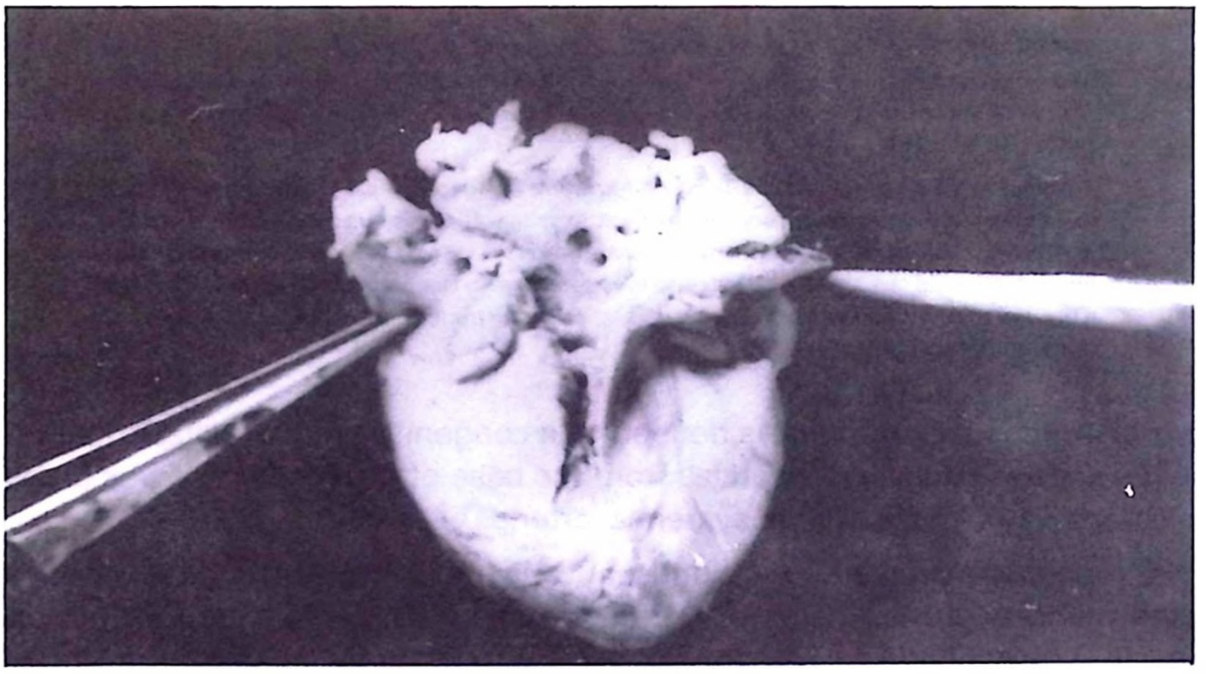

Fig. 1: The general appearance of the heart and hypoplastic left ventricle.

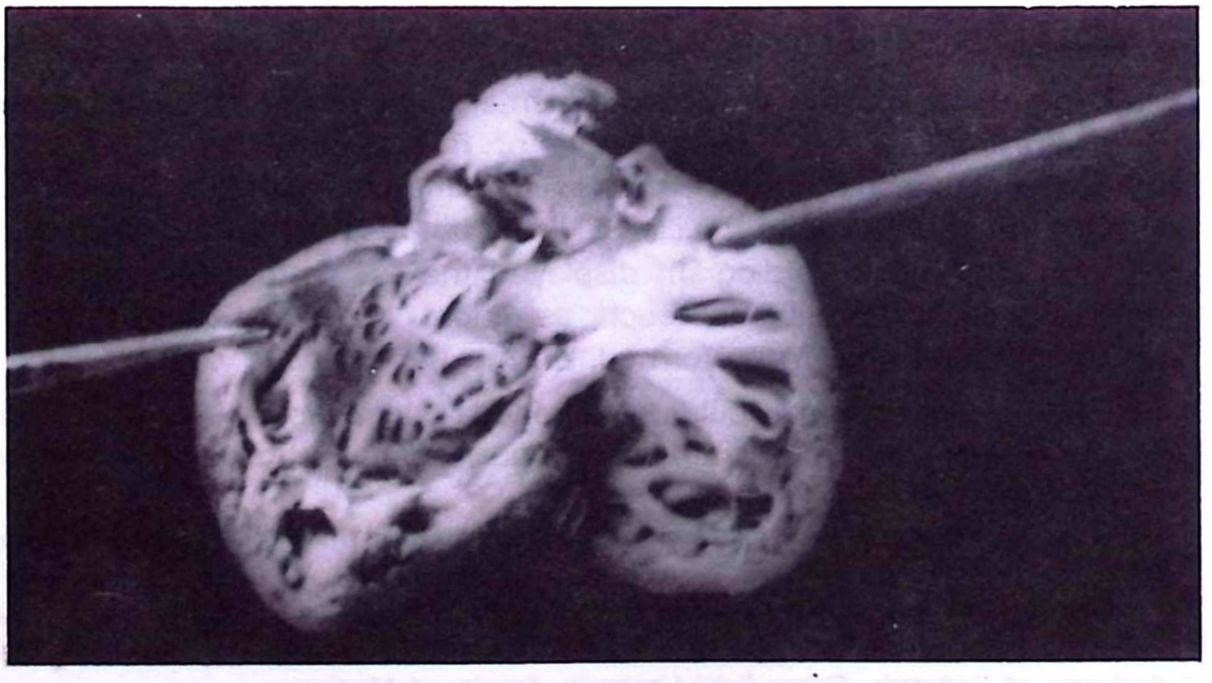

Fig. 2: The enlarged right atrium and the hypertrophied right ventricle.

The atrial and ventricular septa were intact; the small left atrium was draining into the truncus arteriosus at the level of the ascending aorta where the pulmonary arteries left the truncus through an exterior fibrose tunnel-like connection from the postero-inferior wall of the left atrium (Fig. 3).

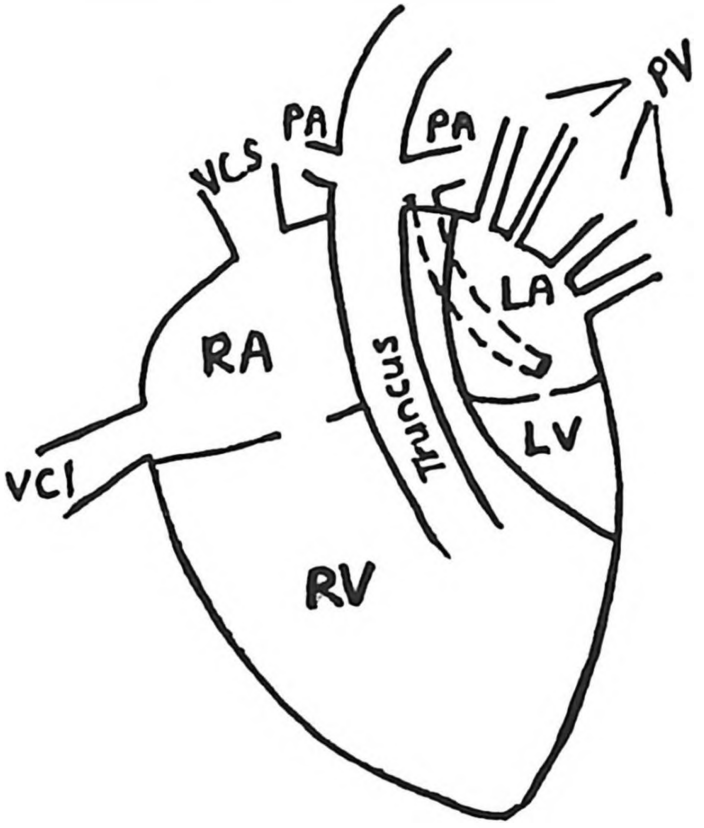

Fig. 3: Demonstration of fibrose tunnel-like connection draining left atrium to the truncus.

Discussion

Truncus arteriosus communis is a rare congenital abnormality, accounting for approximately 1-3% of the largest pathological series of all congenital heart defects and usually accompanied by other intracardiac malformations[3,4]. In our case, truncus arteriosus communis occurred together with hypoplastic left ventricle, intact ventricular and atrial septa, and the left atrium draining to the truncus through a fibrose tunnel-like connection.

No similar case presentation was found, even in the largest series recently reported[2,5,6], endorsing the rarity of this cardiac abnormality. A case of a common arterial trunk arising from the right ventricle with hypoplastic left ventricle, intact ventricular septum and absent left atrioventricular connection was reported by Alves et al in 1987[3], but in their case, there was an atrial septal defect. In our case the atrial septum was also intact and the small left atrium draining into the truncus arteriosus through an exterior fibrose tunnel-like connection was extraordinary. The case is presented because of its unusual nature.

REFERENCES

1. Plauth WH, Nugent EW, Schlant RC, et al. The pathology, abnormal physiology, clinical recognition, and medical and surgical treatment of congenital heart disease. In: Hurst JW, (ed). The Heart, Arteries and Veins. New York: McGraw hill Book Company; 1982: 771.
2. Crupi G, Macartney FJ, Anderson RH. Persistent truncus arteriosus. A study of 66 autopsy cases with special reference to definition and morphogenesis. Am J Cardiol 1977; 40: 569-78.
3. Alves PM, Ferrari AH. Common arterial trunk arising exclusively from the right ventricle with hypoplastic left ventricle and intact ventricular septum. Int J Cardiol 1987; 16: 99-102.
4. Hoffman JI. Congenital heart disease: incidence and inheritance. Pediatr Clin North Am 1990; 37: 25-43.
5. Butto F, Lucas RV Jr, Edwards JE. Persistent truncus arteriosus: pathologic anatomy in 54 cases. Pediatr Cardiol 1986; 7: 95-101.
6. Calder L, Van Praagh R, Van Praagh S, et al. Truncus arteriosus communis. Clinical, angiocardiographic, and pathologic findings in 100 patients. Am Heart J 1976; 92: 23-38.