

Common arterial trunk with intact septum and hypoplastic right ventricle: An uncommon embryological entity

Kalyanraddi Chikkagoudar, Palak Gupta, Nageswara Rao Koneti, Tapan K Dash¹, Vinoth Doraiswamy

Department of Pediatric Cardiology, CARE Hospitals, Hyderabad, Telangana, India, ¹Department of Pediatric Cardiac Surgery, CARE Hospitals, Hyderabad, Telangana, India

ABSTRACT

Embryologically, the common arterial trunk (CAT) is due to the failure of septation by the major outflow cushions. This invariably leads to the presence of an associated large juxta-arterial ventricular septal defect. Overriding of the ventricular septum leads to its biventricular origin in more than 2/3rd of cases. Uncommonly, CAT may be associated with an intact ventricular septum (IVS) and univentricular origin. We describe an infant with CAT, intact IVS, hypoplastic right ventricle, and Ebstein's anomaly of the tricuspid valve who presented to us and palliated successfully. The embryological mechanisms are discussed in this report.

Keywords: Common arterial trunk, Ebstein's anomaly, hypoplastic right ventricle, intact ventricular septum

INTRODUCTION

The common arterial trunk (CAT) can be defined as a single arterial trunk that leaves the heart through a single semilunar valve and gives rise to the coronary, systemic, and one or both pulmonary arteries.^[1] Earlier classifications of CAT were based on the origin of pulmonary arteries, presence/absence of interruption of the arch, and ventricular septal defect.^[2,3] We report an unusual variant of CAT with intact ventricular septum (IVS), Ebstein's anomaly of the tricuspid valve, and hypoplastic right ventricle (RV) who underwent palliative surgery.

CASE REPORT

A 6-month-old male born to nonconsanguineous parents presented with heart failure and cyanosis. The general examination showed dysmorphic features, bounding pulse, and SpO₂ of 82%. Cardiovascular examination revealed hyperdynamic apex, single second heart sound, ejection click, and a 3/6 mid-systolic murmur heard

at the left upper sternal border. Chest X-ray showed cardiomegaly with plethoric lung fields.

Transthoracic echocardiogram (TTE) showed normal situs, CAT with aortic dominance, and intact IVS. The common trunk was committed to the left ventricle (LV) with mitral-semilunar continuity. The RV was hypoplastic (bipartite) associated with Ebstein's anomaly of the tricuspid valve with low-pressure severe tricuspid regurgitation [Figure 1a-d]. The truncal valve was quadricuspid. There was a moderate interatrial communication with obligatory right-to-left shunt. The aortic arch was right-sided with aberrant left subclavian artery. Cardiac catheterization showed net effective left-to-right shunt (Qp/Qs) of 5.1. The right ventricular pressure wave showed atrialized pattern. The calculated Pulmonary Vascular Resistance Index was 3.5 wood units/m². Ventricular and truncal root angiograms essentially demonstrated similar findings [Figure 2a-d].

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Address for correspondence: Dr. Vinoth Doraiswamy, Department of Pediatric Cardiology, CARE Hospitals, Road No. 1, Banjara Hills, Hyderabad - 500 034, Telangana, India. E-mail: dr.vinoth.d@gmail.com

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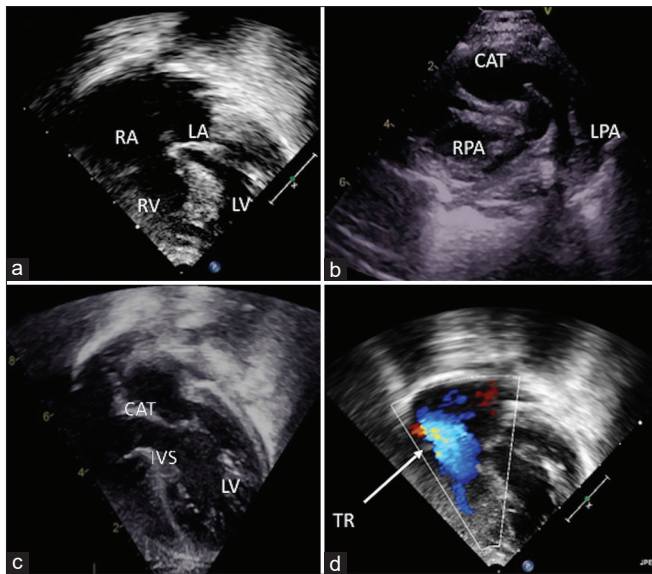


Figure 1: Transthoracic echocardiographic images showing (a) displaced, plastered septal tricuspid leaflet, dilated RA, and hypoplastic functional RV. (b) Parasternal short-axis view demonstrating CAT and both left and right PA. (c) Apical five-chamber view demonstrating CAT arising from LV and intact interventricular septum. (d) Color Doppler in apical four-chamber view, white arrow showing low-pressure severe TR. RA: Right atrium; RV: Right ventricle; PA: Pulmonary artery; CAT: Common arterial trunk; LA: Left atrium; LV: Left ventricle; IVS: Interventricular septum; TR: Tricuspid regurgitation

The only option was a palliative bilateral pulmonary artery banding to prepare for univentricular path. In view of hypoplastic functional RV, the patient underwent bilateral pulmonary artery banding with “*modified Starnes procedure*.” Intraoperative findings confirmed the diagnosis [Figure 3]. Follow-up TTE after 1 month showed well-positioned bilateral pulmonary artery bands with gradient >60 mmHg. He was lost to follow-up and presented at 2 years of age. Cardiac catheterization revealed that he was not suitable for bidirectional cavopulmonary shunt and is currently on medical follow-up.

DISCUSSION

The earlier accepted embryological hypothesis of CAT was the failure of development and fusion of truncoconal cushions, leading to the maldevelopment of three components, namely aortopulmonary septum, truncal septum, and infundibular septum.^[1,2] But now it is clear that, lack of septation of the outflow tracts and intrapericardial arterial roots due to failure of fusion of the major outflow cushions in fetal life is the cause.^[4,6] It is invariably associated with a large juxta-arterial communication due to the failure of fusion of the proximal outflow cushions with the crest of the muscular septum.^[5,6] Only a handful of case reports of CAT with intact IVS have been published,^[7-9] but the embryological explanation in these case reports is not in accordance with what is now known about cardiac development of the outflow tracts.^[5,6]

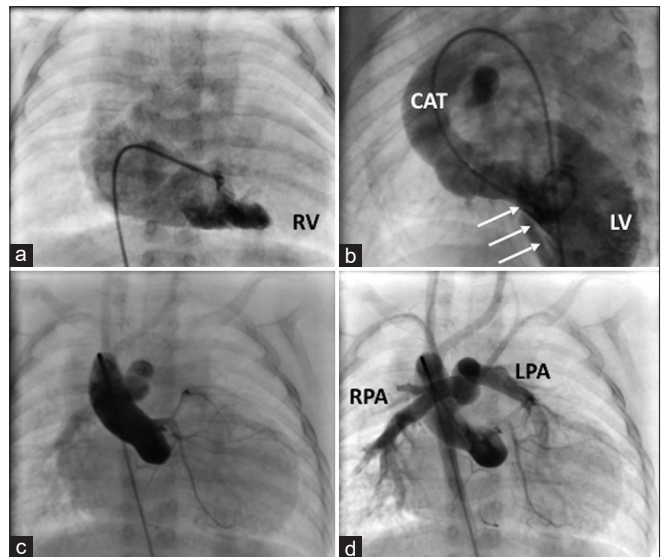


Figure 2: (a) RV angiogram with hand injection in the anteroposterior view demonstrating hypoplastic functional RV and dilated RA due to TR. (b) LV angiogram in LAO cranial view showing the CAT with intact IVS (small multiple arrows). (c) Truncal root angiogram in the anteroposterior view showing a single coronary artery. (d) Truncal root angiogram in the anteroposterior view showing CAT giving rise to both right and left PA, right aortic arch with aberrant left subclavian artery. RV: Right ventricle; PA: Pulmonary artery; CAT: Common arterial trunk; LV: Left ventricle

Embryologically, our case can best be described as failure of fusion of the proximal and distal outflow cushions causing failure of septation of the outflow tracts and intrapericardial arterial roots. Exaggerated commitment of the CAT to LV is followed by the fusion of the proximal outflow cushions with the apical muscular septum. The persisting embryonic interventricular communication gets closed by extensions from the right atrioventricular cushion thus forming the membranous septum and the IVS is intact. Truncal-mitral valve fibrous continuity is produced by the regression of musculature from the inner heart curve. Van Praagh included a type B in his classification consisting of CAT with intact IVS.^[2] But the cases he reviewed had two semilunar valves with intact IVS. These were intact cases of aortopulmonary window caused due to failure of closure of the embryonic aortopulmonary foramen by the dorsal protrusion from the ventral wall of the aortic sac with the distal outflow cushions.^[5,6] These patients have normal outflow cushions which undergo fusion, and hence, two normal semilunar valves, intrapericardial arterial roots, proximal outflow tracts, and an intact IVS. It has recently been proposed to avoid confusion by using alphanumeric classifications^[2,3] and focus on the concept of classifying CAT by aortic dominance/pulmonary dominance/balanced CAT.^[10] Our patient would be classified as aortic dominance as the intrapericardial aorta and aortic arch are not hypoplastic, and there is the separate origin of the pulmonary arteries. This is due to the failure of fusion of ventral protrusion from the dorsal wall of the aortic sac with the distal

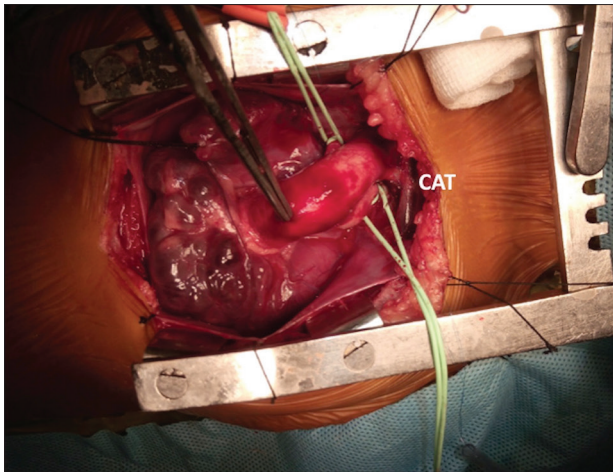


Figure 3: Intraoperative picture showing CAT. The right and left pulmonary arteries are looped. CAT: Common arterial trunk

outflow cushions. This was previously referred to as “aortopulmonary septum” by Van Mierop *et al.*^[4] The association of Ebstein’s anomaly of the tricuspid valve, CAT with intact IVS arising from the left ventricle, and right aortic arch with aberrant left subclavian artery indicates multiple embryological mechanisms.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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