CASE REPORTS

Rare variant of truncus arteriosus with intact ventricular septum and hypoplastic right ventricle

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Abstract

A three week old girl was admitted to hospital with severe congestive heart failure and cyanosis. Cross sectional and Doppler echocardiography and cardiac catheterisation showed a unique variant of truncus arteriosus with an intact ventricular septum. The trunk rose only from the left ventricle and was associated with a hypoplastic right ventricle with sinusoids to the right coronary artery.

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Truncus arteriosus is defined as a single arterial trunk leaving the base of the heart via a single arterial valve to supply the systemic, pulmonary, and coronary circulations.¹ A truncus arteriosus with an intact ventricular septum is rare.² We describe a most unusual case of truncus arteriosus with intact ventricular septum and hypoplastic right ventricle with sinusoids.

Case report

A three week old girl was transferred to our hospital for cardiac evaluation. She was mildly cyanosed with a respiratory rate of 80 per minute and a pulse rate of 150 beats/minute. Blood pressure was 70/40 mm Hg with bounding peripheral pulses. Examination

showed an active precordium with a loud systolic ejection click at the apex. The second heart sound was loud and single. A grade 3/6 ejection systolic murmur was best heard at the apex and the left sternal border. A 2/6 mid-diastolic murmur was best heard at the apex.

A chest x ray showed severe cardiomegaly with increased pulmonary vascular markings. An electrocardiogram showed right atrial enlargement, left ventricular hypertrophy, and diffuse ST-T segment depression.

Cross sectional and Doppler echocardiography showed situs solitus and normal systemic and pulmonary venous connections. The right atrium was very large with aneurysmal bulging of the interatrial septum into the left atrium. The tricuspid valve was hypoplastic but patent and opened into a hypoplastic muscular right ventricle with sinusoids (fig 1A). The mitral valve looked abnormal with moderate stenosis and insufficiency. The left ventricle was large with normal function and the interventricular septum seemed to bulge from right to left in systole. A single arterial trunk originated from the left ventricle: it supplied a left aortic arch, both pulmonary arteries, and the coronary arteries (figs 1B and C). The truncal valve was bicuspid.

Cardiac catheterisation (table) showed a right-to-left shunt at the atrial level. Oxygen saturation was identical in the left pulmonary artery and aorta. The filling pressure of both ventricles was increased. The right ventricular

Figure 1 (A) Apical five chamber view showing the trunclal root (in relation to the left ventricle) and a small hypertrophic right ventricle. (B) Parasagittal suprasternal view of the trunclal arch: the pulmonary artery comes off its posterior aspect. (C) High short axis view of the truncus showing the origin of the pulmonary arteries. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; T, trunclal outlet; V, trunclal valve; PA, pulmonary artery; rp, right pulmonary artery; lp, left pulmonary artery.

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Data from cardiac catheterisation

Site	Oxygen saturation (%)	Pressure (mm Hg)
Superior vena cava	30	—
Right atrium	47	a = 28, v = 26, m = 18
Right ventricle	44	95/18
Left atrium	73	a = 20, v = 28, m = 18
Left ventricle	80	75/22
Aorta	73	60/40, m = 50
Left pulmonary artery	73	55/35, m = 45

pressure was suprasystemic and there was a 15 mm Hg peak systolic gradient across the truncal valve.

CINEANGIOGRAMS

An anterograde injection of contrast into the root of the great artery, which arose from the left ventricle, showed that it supplied the left coronary artery, a left aortic arch, and both pulmonary arteries that rose from its posterior aspect (fig 2A). Though there was good opacification of the right cusp there was no filling of the right coronary artery. There was mild truncal regurgitation. A left ventriculogram showed large ventricle with normal function and mild mitral regurgitation. From this ventricle there was opacification of the truncal artery. A right ventriculogram showed a hypoplastic right ventricle with severe tricuspid regurgitation. From the right ventricle there was filling of multiple sinuoids with retrograde filling of the right coronary artery without opacification of the truncal root (fig 2B).

After the right ventriculogram was obtained severe bradycardia developed and progressed to complete atrioventricular block and hypotension. A rapid balloon atrial septostomy was performed and a temporary pacemaker was inserted but all attempts at resuscitation were unsuccessful.

On the basis of these data we diagnosed an unusual form of truncus arteriosus. The arterial trunk arose only from the left ventricle: the interventricular septum was intact, and the right ventricle was hypoplastic with

multiple sinusoids and retrograde filling of the right coronary artery, which was probably atretic in its origin. There was mild stenosis and insufficiency of the truncal valve, an abnormal mitral valve, and aneurysm formation of the interatrial septum.

Discussion

The present case is unusual in many aspects and does not conform to the conventional classification of truncus arteriosus.^{3,4} However, the definition of truncus arteriosus as a solitary arterial trunk guarded by a common arterial valve and directly supplying the coronary, pulmonary, and systemic circulations would include this unusual variant.¹

The first unusual feature was the origin of the trunk entirely from the left ventricle. This is a rare anomaly found in only 4–6% of patients with truncus arteriosus.^{3,5} In most patients the truncal valve overrides the ventricular septum and arises in approximately equal proportions from the right ventricle and the left ventricle.

The second distinctive feature was the absence of a ventricular septal defect. The ventricular septal defect in truncus arteriosus is generally large and results from either absence or pronounced deficiency of the infundibular septum.⁶ Very rarely the ventricular septal defect in truncus arteriosus may be small and restrictive or even absent.²

The association of left ventricular origin of the truncus arteriosus and absence of a ventricular septal defect left the right ventricle without an outlet. This anatomical arrangement resembles that in cases of pulmonary valve atresia and intact ventricular septum with hypoplastic right ventricle. In our case, as in these cases,⁷ there were multiple sinusoids that connected the right ventricular cavity to the coronary artery and functioned as the sole outlet for this ventricle. The origin of the right coronary artery from the truncus arteriosus was probably atretic because this vessel did not fill from the truncal root injection and there was no retrograde filling of the truncus arteriosus when the right coronary artery filled retrogradely during the right ventricular angiogram.

Figure 2 (A) Truncal root injection showing the anatomy of the root and the filling of the left coronary artery (arrow). (B) Right ventriculogram showing a severely hypoplastic right ventricle with multiple sinusoids and retrograde filling of the right coronary artery (arrow).

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