Persistent truncus arteriosus: a rare finding in adults

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Persistent truncus arteriosus (PTA) comprises <1% of congenital heart defects. Over 80% of infants with truncus arteriosus die within the first year of life without treatment. We are presenting the images of a 44-year-old man with uncorrected PTA, who presented with symptoms of congestive heart failure.

Physical examination was remarkable for cachexy, central cyanosis, jugular venous congestion, clubbed fingers, and massive oedema of the lower legs. Auscultation revealed a mild diastolic murmur parasternal on the right. Other findings were acute renal failure, a reduced liver function, ascites, and bilateral pleural effusion. Blood pressure was 95/50 mmHg and arterial oxygen saturation was 75%. Blood count showed a haemoglobin of 21 mg/dL and a haematocrit of 66%.

Transthoracic echocardiography revealed massive biventricular hypertrophy, dilated right chambers, and severe tricuspid stenosis (Panel A, left hand). Parasternal axis view showed a large truncus arising from the base of the heart, overriding a non-restrictive ventricular septal defect (VSD) supplied by both ventricles (Panel A, right hand). Magnetic resonance angiography (Panel B) showed the truncus giving rise to the coronary arteries, the pulmonary arteries, and the ascending aorta shortly after origin (Type A1 according to van Praagh classification). Furthermore, the thoracic vasculature was significantly altered. The right-sided aortic arch was undercrossed by a large innominate vein, the brachiocephalic trunk arose from the left side of the arch and gave rise to both the left subclavian and left carotid arteries, and the right subclavian and carotid arteries were leaving the arch separately. The course and appearance of the pulmonary veins were normal.

Owing to fixed pulmonary hypertension, we considered to perform a combined heart–lung transplantation. However, the patient refused surgery. Hence, the patient underwent a percutaneous interventional atrioseptostomy and a tricuspid valvuloplasty in one session. This intervention improved the congestion, and by increasing the preload of the functional single ventricle the cardiac output could be enhanced significantly. The clinical situation improved noticeably and 2 weeks later, the patient could be discharged in good condition. In the follow-up, 3 and 6 month later, he presented in good health.

Panel A. Transthoracic echocardiography: four chamber view (left hand) and parasternal axis (right hand). RA, right atrium; RV, right ventricle; LV, left ventricle; LA, left atrium; TA, truncus arteriosus; PE, pericardial effusion. The white arrow indicates the VSD.

Panel B. Volume-rendered technique display of thoracic contrast-enhanced magnetic resonance angiography. TA, truncus arteriosus; PA, pulmonary artery; Ao, aorta; SCV, superior vena cava; DA, descending aorta; IV, innominate vein; LPA, left pulmonary artery; RPA, right pulmonary artery; PV, pulmonary vein.

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