Two different pentalogies in an adult patient: a pentalogy of Cantrell associated with a pentalogy of Fallot

Kamil Tuluce1*, Cemil Gurgun1, Öguz Yavuzgil1, Naim Ceylan2, and Selcen Yakar Tuluce3

1Department of Cardiology, Ege University Faculty of Medicine, 35100 Izmir, Turkey; 2Department of Radiology, Ege University Faculty of Medicine, Izmir, Turkey; and 3Department of Cardiology, Ataturk Training and Research Hospital, Izmir, Turkey

* Corresponding author. Tel: +90 232 3904001; fax: +90 232 3903287, Email: kamiltuluce@gmail.com

Pentalogy of Cantrell is a rare syndrome characterized by defects involving the abdominal wall, lower sternum, diaphragm, pericardium, as well as congenital cardiac anomalies. Tetralogy of Fallot is a cardiac anomaly consisting of a ventricular septal defect (VSD), dextroposition of the aorta, obstruction to the pulmonary blood flow, and right ventricular (RV) hypertrophy. Addition of atrial septal defect (ASD) to these anomalies is called pentalogy of Fallot (POF).

A 33-year-old woman presented with effort intolerance. She had a history of cardiac surgery 26 years ago. No repair procedures could be performed because of the complex cardiac anomalies detected unexpectedly. Cardiac auscultation revealed a systolic ejec-

tion murmur at the lower left sternal border and the apex. A pulsation simultaneous with pulse was visible in the substernal region. Clubbing and cyanosis were not detected. Transthoracic echocardiography showed an apical outpouching of the left ventricle. (B) Coronary angiography examination showed an anomalous origin of the left anterior descending artery arising from the right aortic sinus, and (C) the origin of the circumflex artery was the left sinus of Valsalva. (D) Computed tomography coronal 3-dimensional image demonstrated left ventricular hipoplasia, and (E) coronal multiplanar reformatted scan revealed an LV diverticulum (short arrow) and anterior diaphragmatic defect (long arrow). (F) Coronary maximum intensity projection magnetic resonance angiography examination image demonstrating interventricular septum (short arrows), VSD (thick arrow), and dextroposition of the aorta (long arrows). Magnetic resonance also visualized a secundum type ASD (G) and stenosis of pulmonary valve (H). RCA, right coronary artery; LAD, left anterior descending artery; CX, circumflex artery.

A 33-year-old woman presented with effort intolerance. She had a history of cardiac surgery 26 years ago. No repair procedures could be performed because of the complex cardiac anomalies detected unexpectedly. Cardiac auscultation revealed a systolic ejec-
tion murmur at the lower left sternal border and the apex. A pulsation simultaneous with pulse was visible in the substernal region. Clubbing and cyanosis were not detected. Transthoracic echocardiography showed an apical outpouching of the left ventricle (LV) diverticulum (Figure 1A). POF, including a VSD with bidirectional shunt, an ASD, pulmonary stenosis, RV hypertrophy, and an overriding aorta, was also visualized. Coronary angiography demonstrated an anomalous origin of the LAD artery arising from the right aortic sinus (Figure 1B and C). Left ventriculography showed an LV diverticulum, a VSD, and a partial thoracoabdominal ectopia cords (Supplementary data online, Video S1). Cardiac MR and thoracic CT showed the anatomical course and the focal bulging of the
diverticulum under the skin (Supplementary data online, Video S2) in the substernal region due to the lack of xiphoid and the defects of anterior part of both the pericardium and the diaphragm (Figure 1D and E; Supplementary data online, Video S3). The presence of POF was also demonstrated (Figure 1F–H). The diagnosis of incomplete Cantrell’s pentalogy was made with cardiac involvement in forms of POF, an LV diverticulum, and a coronary artery origin anomaly. Surgery was planned but the patient refused. She has been followed for 18 months without any events.

**Supplementary material**
Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

**Conflict of interest:** None declared.