CJC Pediatric and Congenital Heart Disease 1 (2022) 198–199

Images in Cardiology

Pentalogy of Cantrell

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Pentalogy of Cantrell (POC) consists of 5 congenital defects that include defects of the heart, pericardium, sternum, diaphragm, and the anterior abdominal wall. The reported incidence varies between 1:65,000 and 1:200,000 births,[1] it is crucial to promptly identify these patients to correct all the associated anomalies. Patients with pentalogy of Cantrell usually have ectopia cordis, a condition where the heart protrudes to the ventral abdominal wall, and it is covered by an omphalocele-type membrane. Multiple cardiac anomalies can be found in these patients, the most common being ventricular septal defects and tetralogy of Fallot.[2]

These are the images of a 28-day-old female neonate, obtained by a caesarean section at the 40 weeks of gestation, as a product of a second pregnancy that had a normal evolution. At the 32 weeks of gestation, an epigastric hernia with ectopia cordis covered by pericardium was observed. Postnatal echocardiography was performed, showing the absence of sternum, a common atrium, single, double outlet, morphologic right ventricle with complete atrioventricular septal defect with a common atrioventricular valve and pulmonic stenosis (maximum gradient of 61 mm Hg) and posterior aorta (Fig. 1A-C). Cardiac magnetic resonance imaging was performed, confirming the echocardiographic findings of bronchial and abdominal situs solitus (Fig. 2A-E).

Because treatment involves an extensive surgical approach, different image modalities are necessary in the preoperative evaluation. Thus, the case highlights the importance of high-quality imaging assessment, which aids the surgical team to adequately plan the intervention.

Novel Teaching Points

- Transthoracic echocardiography continues to be a fundamental tool in the assessment of thoracic anomalies, with good correlation with magnetic resonance imaging (MRI) findings.
- Cardiac MRI helps characterize the anatomy of cardiac and paracardiac structures, allowing adequate visualization of the aorta and thoracic spine.
- Three-dimensional cardiac MRI reconstructions provide high-quality images that aid the surgical team to adequately plan the surgical procedure.

Ethics Statement

The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Funding Sources

No funding was received for this study.

Disclosures

The authors have no conflicts of interest to disclose.

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Received for publication May 23, 2022. Accepted June 10, 2022.

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https://doi.org/10.1016/j.cjcpc.2022.06.003

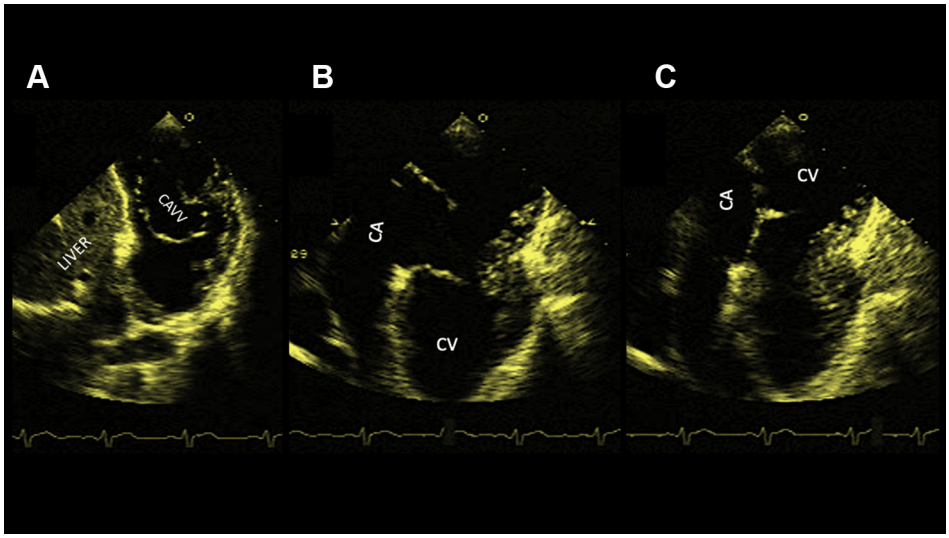

Figure 1. (**A**) Two-dimensional transthoracic echocardiogram in subcostal view showing atrioventricular connection with the common AV valve. (**B, C**) Single morphologic right ventricle with the common AV valve in diastole and systole with double outlet. AV, atrioventricular valve; CA, common atrium; CAVV, common atrioventricular valve; CV, common ventricle.

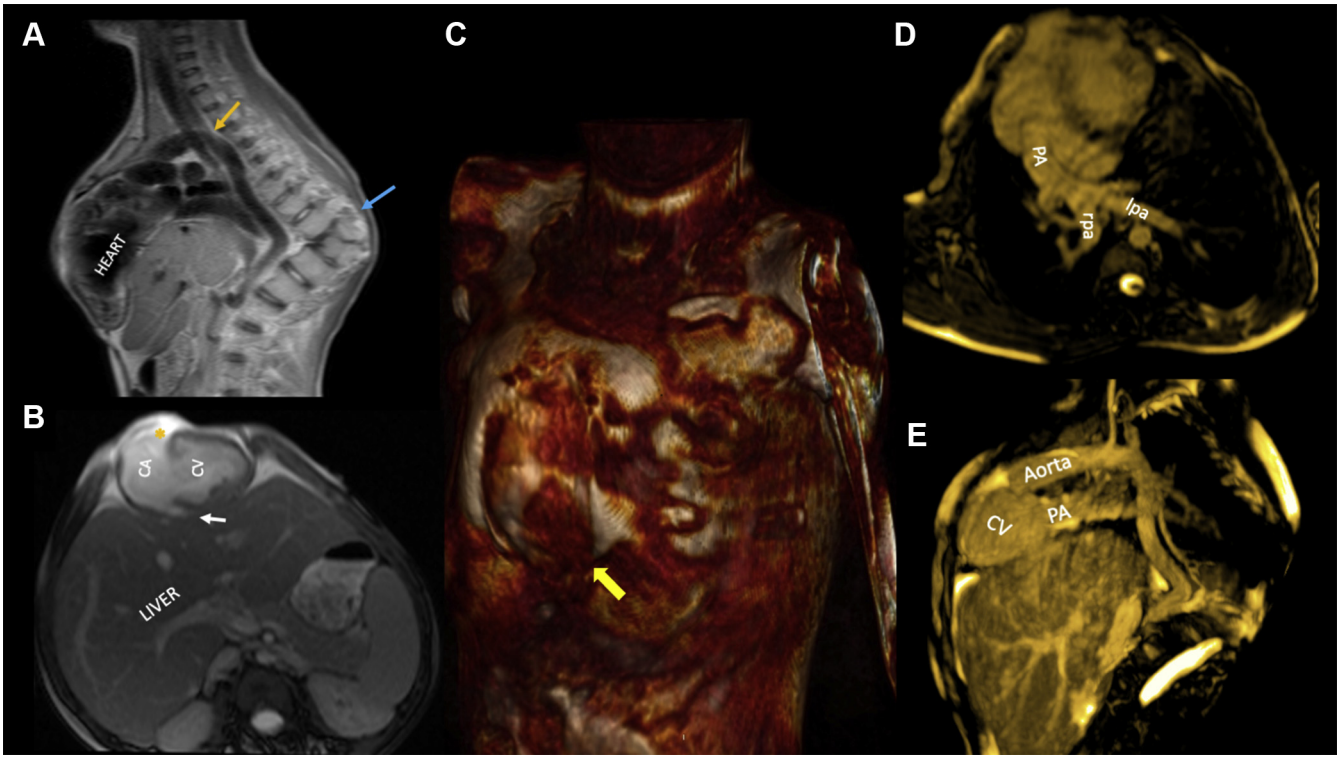

Figure 2. (**A**) Cardiac magnetic resonance. T1-sequence MRI in sagittal view showed tortuosity in the aorta (**yellow arrow**), ectopia cordis and severe kyphosis of the thoracic column (**blue arrow**). (**B**) T2-weighted sequences presenting atrial situs solitus (right and left atrial appendages marked with an **asterisk** and **white arrow**) with a common atrioventricular valve and single ventricle, central liver, and loss of the continuity of the anterior thoracic wall due to the absence of the sternum. (**C**) Three-dimensional (3D) volumetric reconstruction showed a thoracic mass corresponding to the ectopic heart. (**D**) 3D navigator in axial view evidenced the main pulmonary artery and its branches. (**E**) 3D navigator in sagittal view confirmed the tortuosity in the aorta and showed the outlet of the main pulmonary artery, both arising from the single ventricle.CV, common ventricle; LPA, left pulmonary artery; PA, pulmonary artery; RPV, right pulmonary artery.