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Images in Congenital Cardiac Disease

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Abstract

Coronary cameral fistula is a rare congenital anomaly and clinical presentation depends on the location of the defect, degree of shunting, and associated complications. We present a case of coronary cameral fistula where segmental analysis by echocardiogram helped us to avoid misdiagnosis as a ventricular septal defect.

Case: An 8-year-old girl was incidentally detected to have a ventricular septal defect due to a continuous murmur with IV/VI intensity found on physical examination. Electrocardiography showed sinus rhythm and non-specific ST-T changes. Chest X-ray was suggestive of normal pulmonary blood flow with no cardiomegaly. Transthoracic echocardiography showed a red jet into the right ventricle with a defect in the inter-ventricular septum mimicking a muscular ventricular septal defect (Fig. 1a, b, supplementary video 1,2) with mildly dilated left ventricle. However, flow was continuous, noted in both diastole and systole (Fig. 1c). The gradient across the defect was 110/61mmHg (Fig. 1c). The parasternal long-axis view showed a grossly dilated left circumflex artery in the left atrioventricular groove with turbulent flow within (Fig. 1d). Aneurysmal left main origin continuing as a huge serpiginous left circumflex artery in the left atrioventricular groove draining into the right ventricle via inter-ventricular septum can be traced in an echocardiographic sweep (Fig. 2a–h, supplementary video 3). Cardiac contrast computed tomogram confirmed the same (Fig. 3a–e, Fig. 4a–d), and showed left anterior descending, right coronary and obtuse marginal arteries of normal calibre (Fig. 4a–d), and thus coronary cameral fistula involving left circumflex artery and draining to the right ventricle, mimicking a ventricular septal defect. Coronary cameral fistulas are rare cardiac malformations, seen only in 0.002% of the population.¹ The presentation of coronary cameral fistulas varies from asymptomatic at an early age to symptomatic and starts complications upon ageing. Coronary cameral fistulas may be misdiagnosed as ventricular septal defect if the exit is into the right ventricle through the inter-ventricular septum. However, continuous Doppler flow through the defect, the presence of dilated coronaries and no colour jet truly crossing the inter-ventricular septum were suggestive of a coronary cameral fistula in our case. Multimodality imaging (coronary CT angiogram) has an immense role in confirming the diagnosis and in surgical/transcatheter planning in hemodynamically significant cases. Our case emphasises the routine segmental analysis by echocardiogram to avoid misdiagnosis and even to help us reach a rare diagnosis with certainty.

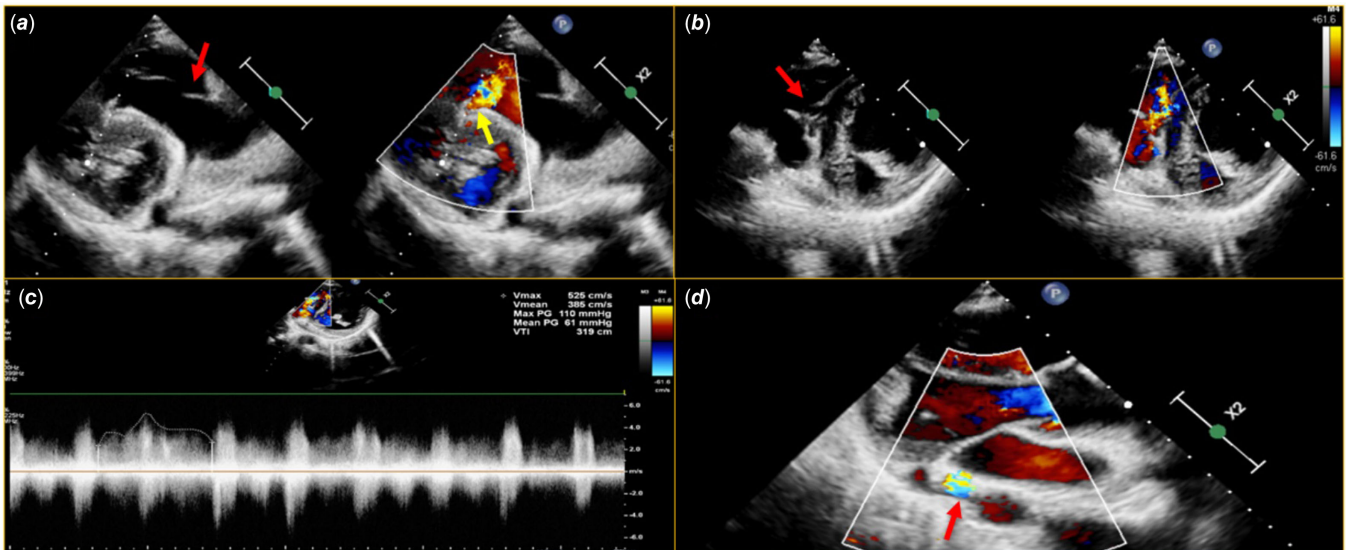


Figure 1. *a.* Modified parasternal long-axis view showing a red jet into right ventricle (yellow arrow), note that flow is in diastole while tricuspid valve is open (red arrow); *b.* Parasternal short axis view showing similar red jet with defect in the inter-ventricular septum (arrow), mimicking a ventricular septal defect; *c.* Gradient across the defect was 110/61mmHg, note the continuous flow; *d.* Parasternal long-axis view showing dilated left circumflex artery in left atrioventricular groove (arrow).

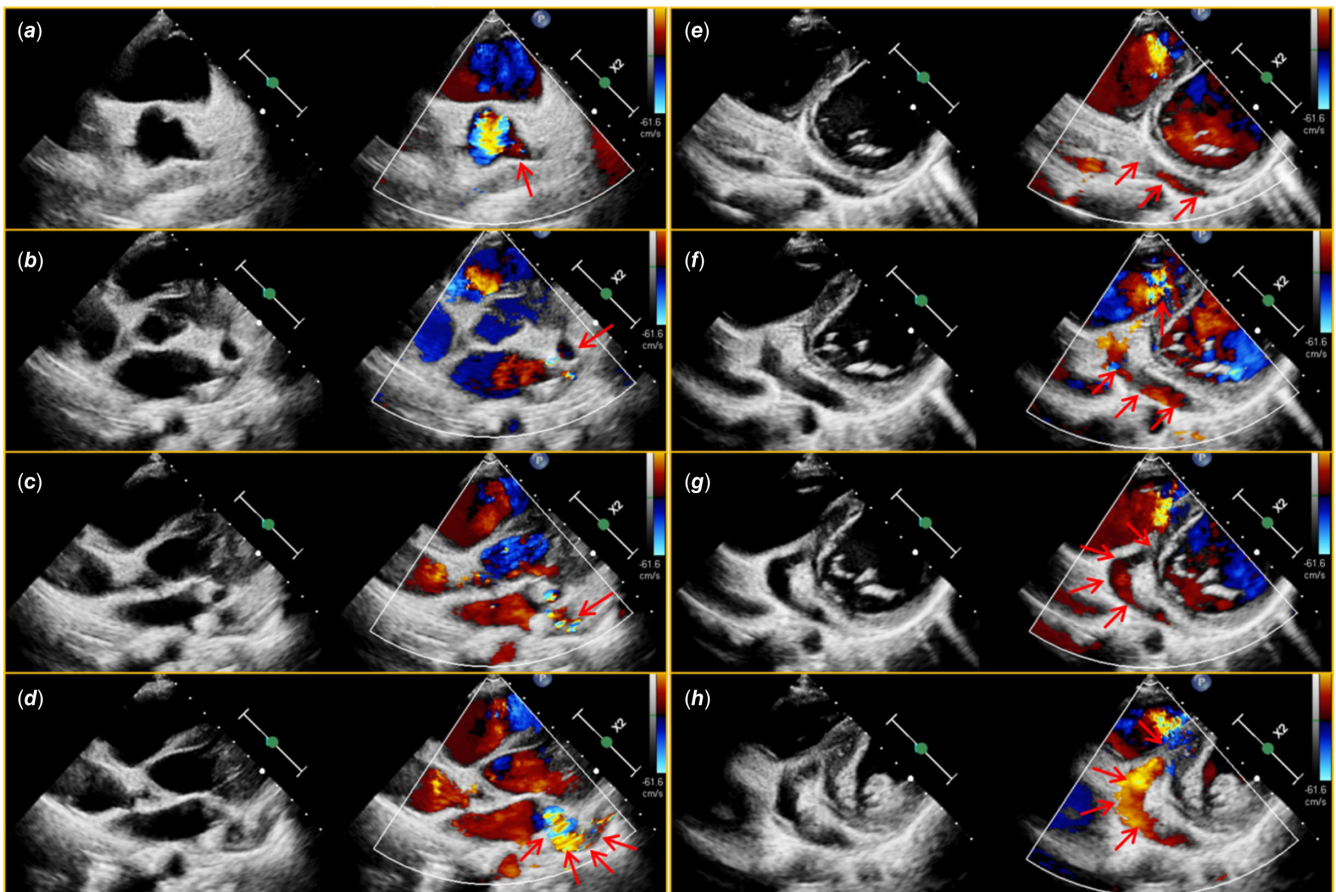


Figure 2. *a-h.* The parasternal short axis sweeps from the base to the mid-ventricular segment tracing the hugely dilated left main (arrow in A) and left circumflex artery (arrows) in the atrioventricular groove, ultimately draining to the right ventricle via an inter-ventricular septum.

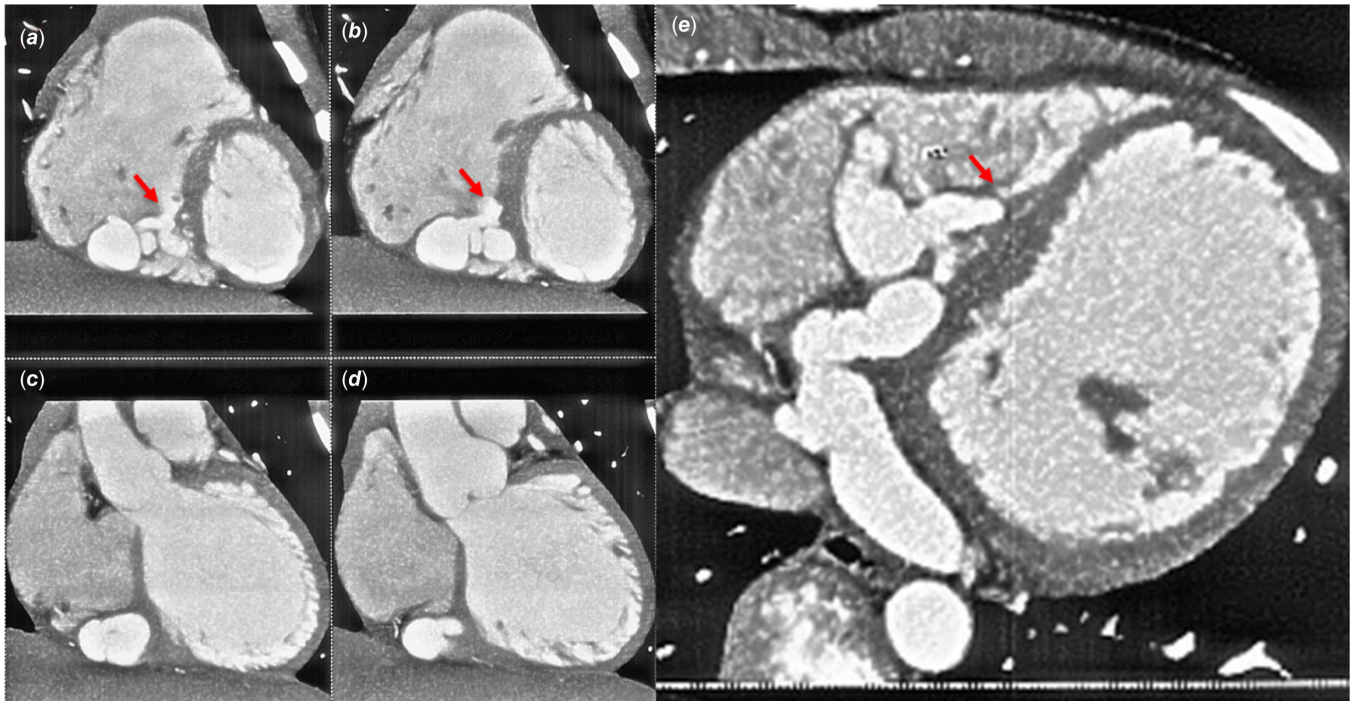


Figure 3. *a–d.* Cardiac contrast CT in coronal section showing hugely dilated left circumflex artery draining (arrow) into right ventricle via inter-ventricular septum; *e.* Axial contrast computed tomogram showing the same (arrow).

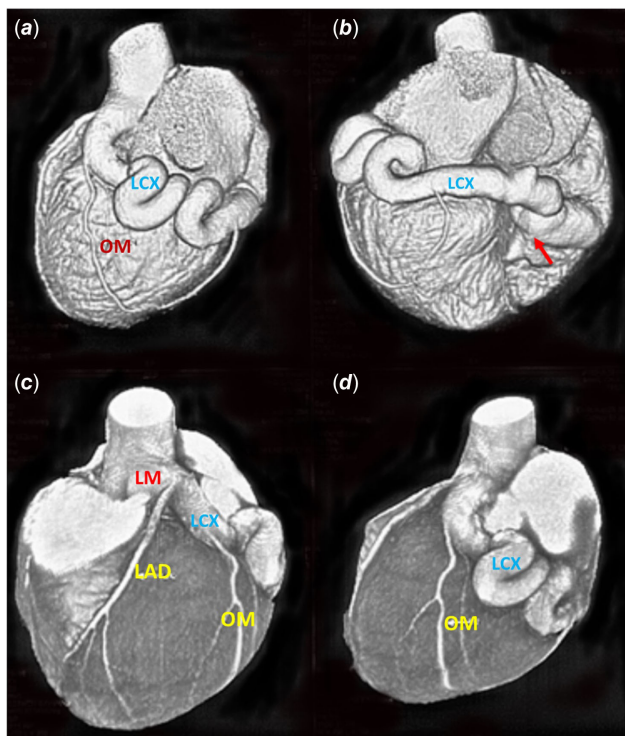


Figure 4. The surface anatomy, course, and shape of the giant tortuous coronary fistula, as well as its anatomic relationships with the LAD, are readily appreciated in the volume-rendering technique images, showing the lateral (*a*), posterior (*b*), left oblique cranial (*c*), and left lateral cranial (*d*) aspects of the heart; the origin and the termination (arrow) site of the coronary fistula are clearly depicted. LAD = left anterior descending artery; LM = left main artery; LCX = left circumflex artery; OM = obtuse marginal artery.

Supplementary material. The supplementary material for this article can be found at <https://doi.org/10.1017/S104795112300389X>.

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Competing interests. None.

Consent. Obtained from the patient in line with COPE guidance.

Reference

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