

# Echocardiographic Diagnosis of Incidentally Found Left Coronary Artery to Pulmonary Artery Fistula in an 11-Year-Old Girl

Bo-Hau Chen,<sup>1,4</sup> Chu-Chuan Lin,<sup>1</sup> Ken-Pen Weng,<sup>1,2,3</sup> Huang-Wei Wu,<sup>5</sup> Jen-Hung Chien<sup>6</sup> and Shih-Ming Huang<sup>7</sup>

We report on a healthy 11-year-old girl who presented to our facility with sudden onset of fainting in a strenuous running course. Transthoracic echocardiography at short-axis view showed a diastolic flow into the main pulmonary artery (PA). The diagnosis of left anterior descending artery (LAD) to PA fistula was documented by cardiac computed tomography and catheterization. Interventional therapy of LAD to the main PA fistula was not performed because of no evidence of myocardial ischemia or significant hemodynamic change. Presently, the patient remains asymptomatic. Coronary fistula with an incidence of about 0.1-0.8% is very rare and may be undetected, particularly in pediatric patients without cardiac murmur. We herein describe the diagnostic approach and discuss the current treatment modalities.

**Key Words:** Coronary angiography • Coronary artery fistula • Echocardiography

## INTRODUCTION

Coronary artery fistula (CAF) is rare and may be congenital or acquired.<sup>1</sup> In Taiwan, CAF has a prevalence rate of less than 1%, and is usually asymptomatic.<sup>2</sup> It may be found incidentally with coronary angiograms or cardiovascular computed tomography (CT).<sup>2,3</sup> However, transthoracic echocardiography (TTE) can be used to diagnose most of the cases and is a better noninvasive screening tool to detect coronary artery fistula in pediatric patients than coronary angiograms or CT in terms

of expense and radiation.<sup>3,4</sup> We report a case with left anterior descending artery (LAD) to the main pulmonary artery (PA) fistula in an 11-year-old girl, which was initially suspected on routine TTE examination.

## CASE REPORT

An 11-year-old girl was in generally good health except for mild intermittent dyspnea during strenuous exercise. She suffered from a sudden onset of fainting during a course of strenuous running before admission to our hospital. Physical examination revealed irregular heart beats and slight diastolic murmur with a relatively strong peak sound mimicking pulmonary insufficiency. The patient's chest X-ray, hematology, biochemical indices and serial cardiac enzymes were all unremarkable apart from mild raised brain natriuretic peptide (BNP) levels of 40.1 pg/mL. Electrocardiography (ECG) at our emergency room demonstrated non-specific ST-T elevation in V<sub>2</sub>-V<sub>6</sub> (Figure 1A). On admission, TTE at short-axis view showed a diastolic abnormal color Doppler flow into the main PA (MPA) (Figures 2A, 2B, and 2C). Left

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<sup>1</sup>Department of Pediatrics, Kaohsiung Veterans General Hospital, Kaohsiung; <sup>2</sup>National Yang-Ming University, Taipei; <sup>3</sup>Shu-Zen College of Medicine and Management, Kaohsiung; <sup>4</sup>Department of Pediatrics, Taoyuan Armed Forces General Hospital, Taoyuan; <sup>5</sup>Department of Pediatrics, Kaohsiung Armed Forces General Hospital; <sup>6</sup>Department of Pediatrics, Zuoying Branch of Kaohsiung Armed Forces General Hospital; <sup>7</sup>Department of Pediatrics, Kaohsiung Municipal United Hospital, Kaohsiung, Taiwan.

Address correspondence and reprint requests to: Dr. Ken-Pen Weng, Department of Pediatrics, Kaohsiung Veterans General Hospital, No. 386, Ta-Chung 1<sup>st</sup> Road, Kaohsiung 813, Taiwan. Tel: 886-7-346-8202; Fax: 886-7-346-8207; E-mail: kenpenweng@yahoo.com.tw

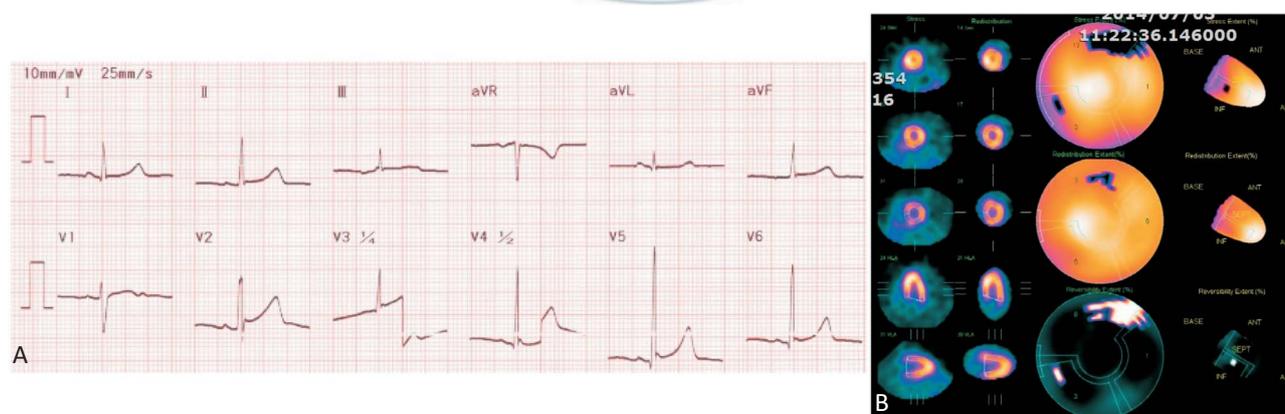
coronary artery to the MPA fistula was diagnosed after careful and repeated short-axis view. The diagnosis of LAD to MPA fistula was documented by CT coronary angiography (Figures 2D and 2E). Nuclear cardiac examination of ejection fraction plus wall motion showed preserved biventricular systolic function and preserved left ventricle diastolic function. Dipyrimadole myocardial perfusion scan showed mild reversible defect in the anterior wall (30% decrease) (Figure 1B). Cardiac catheterization was performed for hemodynamic study and possible intervention occlusion. The patient's left coronary angiography showed a tube-like fistula originating from the proximal LAD emptying into PA in the anteropostero-caudal view (Figure 2F), but multiple small fistulas in the lateral-caudal view (Figure 2G). Cardiac catheterization showed the pulmonary-to-systemic flow ratio ( $Q_p/Q_s$ ) = 1.0 and mean PA pressure = 21 mmHg, respectively. Transcatheter embolization or surgical intervention was not indicated due to the lack of evidence of myocardial ischemia or significant hemodynamic change ( $Q_p/Q_s$  = 1.0 and mean PA pressure = 21 mmHg). She was asymptomatic at the outpatient department follow-up and was recommended to avoid strenuous exercise.

## DISCUSSION

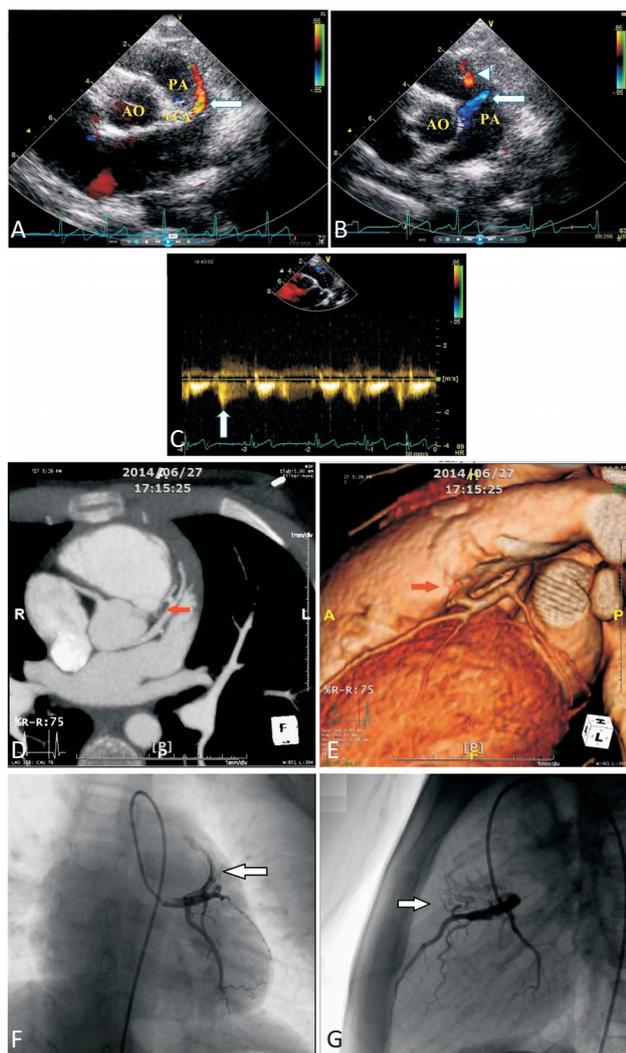
Congenital CAF is found in 0.1-0.8% of patients who undergo coronary angiography.<sup>2,5</sup> The majority of these fistulas originate from the right or left anterior descend-

ing coronary artery.<sup>2-5</sup> Low-pressure structures, such as pulmonary artery, are the most common sites of drainage of the coronary fistula.<sup>5</sup> Most patients are asymptomatic, but symptoms of heart failure, pulmonary hypertension, coronary ischemia or even myocardial infarction, and endocarditis may occur in some cases with significant shunts as patients increase in age.<sup>1</sup> Pediatric patients are usually identified because of electrocardiographic or chest X-ray abnormality, or a loud continuous cardiac murmur.<sup>6</sup> CAF may be undetected in asymptomatic patients without heart murmur.<sup>1</sup> The importance of early identification of anomalous origin of coronary artery to prevent sudden death is well-known, but most asymptomatic pediatric patients with small CAFs diagnosed during routine echocardiography may be managed conservatively and require no further image studies. Because our patient presented with dyspnea and sudden onset of fainting during the strenuous exercise, the diagnosis of a small LAD to PA fistula is still significant clinically and merits further evaluation.

In this case, physical examination revealed a slight diastolic murmur with a relatively strong peak sound mimicking pulmonary insufficiency. The diastolic murmur is caused by diastolic flow of CAF into PA, as demonstrated in our echocardiography. Although there is no evidence of myocardial ischemia, EKG at our emergency room showed non-specific ST-T change in V2-V6, which is compatible with the finding of suspected mild reversible defect in the anterior wall (30% decrease) and mild ischemia in LAD territory by persantin myocardial perfusion scan. The diastolic murmur and nonspecific ST-T



**Figure 1.** (A) Electrocardiogram showed non-specific ST elevation in V2-V6. (B) Hot apical uptake of persantin myocardial perfusion scan showed mild reversible defect in anterior wall (30% decrease).



**Figure 2.** (A) Color Doppler from parasternal short axis view showed an abnormal blood flow (white arrow) within the left coronary and (B) simultaneous appearance of pulmonary regurgitation (white arrowhead) and diastolic fistula flow (white arrow) in the MPA. (C) Continuous-wave Doppler from parasternal short axis view showed diastolic fistula flow with a peak jet (white arrow) mimicking pulmonary insufficiency. (D) Cross-sectional computed tomography image showing contrast-enhanced blood vessels (red arrow) without dilatation connecting proximal LAD and MPA. (E) Three-dimensional image of heart revealed coronary arteriovenous fistulae located from LAD to MPA (red arrow). (F) Anteroposterio-caudal view of left coronary angiogram showing one tube-like fistula (white arrow) originating from proximal LAD to MPA. (G) Lateral-caudal view showing numerous micro-fistulas (white arrow) from proximal LAD to MPA. AO, aorta; LAD, left anterior descending artery; MPA, main pulmonary artery; PA, pulmonary artery.

change in V2-V6 may suggest two clinical clues of cardiac disease. Echocardiography has a high value in showing the origin of CAF. The important hint indicative of CAF is detection of abnormal Doppler diastolic flow of

PA. Differential diagnosis includes persistent ductus arteriosus, coronary artery aneurysm, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect, internal mammary artery to pulmonary artery fistula, and systemic arteriovenous fistula.<sup>5</sup> Cautious and experienced use of echocardiography is very important in avoiding a misdiagnosis. Compared to echocardiography, 64-MSCT angiography<sup>3</sup> can provide more detailed anatomical images for further intervention, as demonstrated in our case.

Possible therapeutic options include surgical correction and transcatheter embolization.<sup>1</sup> The main indications for closure are clinical symptoms including heart failure and myocardial ischemia, and in asymptomatic patients with significant shunt ( $Q_p/Q_s > 1.5$ ) to prevent occurrence of symptoms or complications, especially in a pediatric population.<sup>1,7,8</sup> Regular follow-up is recommended in asymptomatic patients.<sup>1,5</sup> Antiplatelet or anticoagulant therapy is indicated in patients with coronary stenosis or dilatation, which may lead to potential risks of thrombotic events and myocardial ischemia.<sup>1</sup> The possibility of spontaneous closure has been reported.<sup>9</sup> Catheterization in our case showed multiple small fistulous of LAD to PA, and transcatheter embolization or surgical intervention was not performed because of no evidence of myocardial ischemia or significant hemodynamic change. Incorporating the previous data and the catheterization results from our case, using the “wait and see” approach<sup>10</sup> to her medical management may be well-advised. However, there is a sub-clinical effect of CAF in this patient, especially during strenuous exercise, so avoidance of strenuous exercise is recommended.

In conclusion, echocardiography is useful in the diagnosis of CAF, especially in children. However, the treatment of incidentally-found and asymptomatic CAF in children remains controversial, and regular follow-up is recommended.

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