A 7-year-old girl was brought to the pediatric outpatient clinic because of exertional dyspnea for 2 years and chest pain for 2 months. Physical examination showed grade 2/6 systolic murmur at left lower sternal border. Electrocardiogram (ECG) revealed left ventricular hypertrophy with left axis deviation. Transthoracic echocardiogram showed enlargement of the left atrium and moderate mitral regurgitation. Then, she was referred to cardiovascular magnetic resonance (CMR) for potential coronary anomaly. Coronary MRA was performed using ECG-gated, respiratory-navigated inversion-recovery prepared 3D-segmented gradient echo sequence with infusion of contrast agent. CMR demonstrated left coronary artery (LCA) originating from inferior aspect of pulmonary trunk (Figure 1A). The dilated right coronary artery arising normally from the right sinus of Valsava branched into posterior descending artery. The dynamic contrast-enhanced perfusion myocardial imaging showed septal subendocardial perfusion defect persisting for eight frames (Figure 1B). Mitral regurgitation was also clearly depicted in cine gradient echo image. The diagnosis of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) was established.

She received direct aortic reimplantation surgery to correct coronary artery circulation. Follow-up CMR after surgery showed that the anomalous LCA was reimplanted to aortic root successfully with patency.

ALCAPA was first described in 1866, but the full clinical-anatomic description was given by Bland, White, and Garland in 1933, so the anomaly is also called Bland–White–Garland syndrome. ALCAPA is a rare and life-threatening congenital coronary artery anomaly. Reversed LCA flow into pulmonary artery caused by decreased pulmonary artery pressure after birth can lead to myocardial ischemia, left ventricular failure, and sudden death. The management of ALCAPA includes surgical reimplantation of the coronary artery to the aorta.
dysfunction, ischemic mitral regurgitation and sudden cardiac death. Like this case, a child with echocardiographic diagnosis of significant mitral regurgitation who has no congenital valve anomaly and previous history of rheumatic heart disease should prompt the search of ALCAPA.

Conventionally, ALCAPA is diagnosed by conventional angiography, which is now replaced by advanced cardiac CT and CMR. CMR is superior to cardiac CT for pediatric patients due to no radiation, no requirement of breath hold and no need to lower heart rate. In addition, perfusion CMR has superior spatial resolution than single-photon emission computed tomography in detecting subendocardial ischemia, which is important for postoperative prognosis.

CMR can provide all-in-one assessment of the coronary anatomy, myocardial ischemia and ventricular-valvular function of ALCAPA in a pediatric patient with unexplained mitral regurgitation. Reimplantation of LCA is the definite treatment and has excellent results.

Photographs and text from: H.-T. Hsu, Department of Radiology, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan; M.-T. Wu, Department of Radiology, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan and School of Medicine, National Yang-Ming University, Taipei, Taiwan. email: rondavm3@gmail.com

Conflict of interest: None declared.

References