



## Case Study

# An extremely rare case of anomalous origin of the left coronary artery from the pulmonary artery concomitant with noncompaction of the ventricular myocardium in a half marathon runner

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**Keng-Yi Wu,<sup>1,2,3</sup>  
and  
Shu-Meng Cheng\*<sup>2</sup>**

<sup>1</sup>Department of Cardiology,  
Department of Internal Medicine,  
Taichung Armed-Forces General  
Hospital, Taichung, Taiwan,  
Republic of China.

<sup>2</sup>Division of Cardiology,  
Department of Internal Medicine,  
Tri-Service General Hospital,  
National Defense Medical Center,  
Taipei, Taiwan, Republic of China.

<sup>3</sup>Graduate Institute of  
Radiological Science, Central  
Taiwan University of Science and  
Technology, Takun, Taichung 406,  
Taiwan; Republic of China.

\*Corresponding Author E-mail  
[magidino520@yahoo.com.tw](mailto:magidino520@yahoo.com.tw)

Tel: +886-2-8792-7160

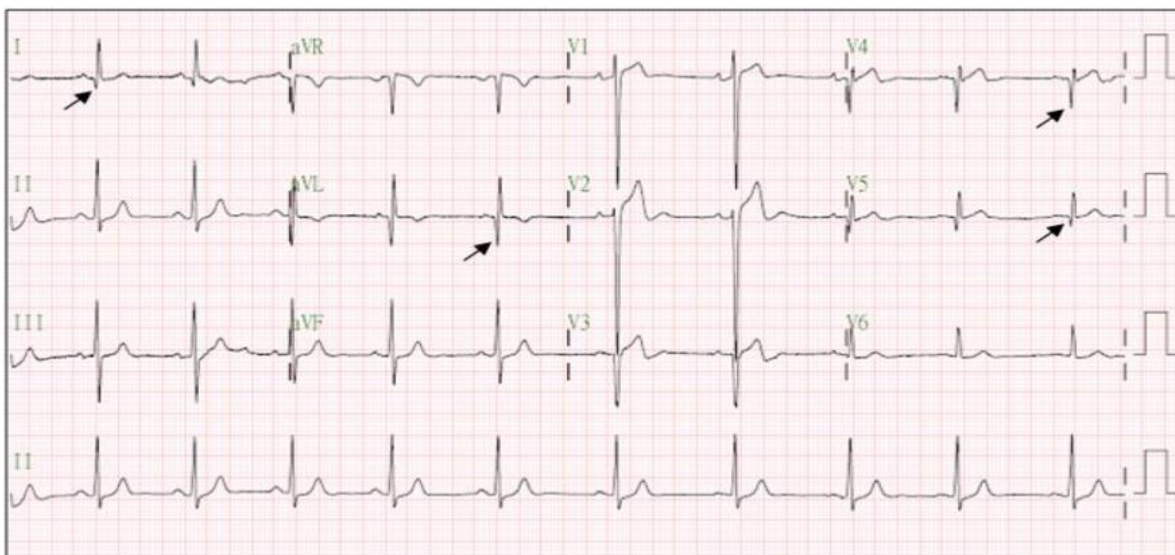
**Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare coronary anomaly disease in the world. Noncompaction of the left ventricular myocardium (LVNC) is also another rare disease in cardiovascular field. With the progressive clinical symptoms, these patients can not do excessively intense, such as marathon or swimming. Bypass surgery with optimized medications is a good chosen therapy initially. Cardiac transplantation is an actual cured treatment for these patients when they reach the end stage heart failure. Here we report a 22-year-old half-marathon man runner who was survived cardiac arrest secondary to undetected anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) concomitant with LVNC.**

**Keywords:** Anomalous origin of the left coronary artery from the pulmonary artery, noncompaction of the ventricular myocardium, marathon player, cardiac arrest.

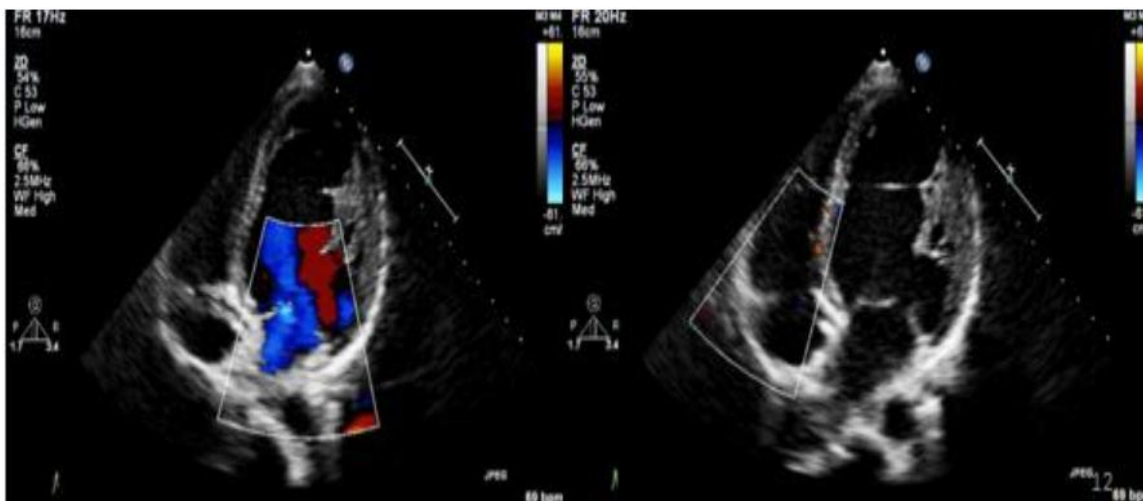
## INTRODUCTION

Coronary artery anomalies can present as origin, termination or numbers abnormality. These diseases are found around in 0.25% to 1% of the population (Yamanaka and Hobbs, 1990). Some of the disease was be found accidentally by electrocardiogram (ECG) or Computed Tomography (CT) for different diagnoses. ALCAPA is a very rare coronary artery anomaly in general

population. Clinical symptoms could present as angina, dyspnea, syncope, arrhythmia or even cardiac arrest (Alsara et al., 2014). LVNC is also a rare disease which prevalence rate about 0.05% in adults. LVNC can be detect by echocardiography or cardiac magnetic resonance imaging (MRI) (Liu et al. 2018; Zhu et al., 2018). Clinical presentations are shortness of breath,



**Figure 1:** Electrocardiogram. Pathological Q wave (arrow)



**Figure 2:** Prominent ventricular trabeculations with deep intertrabecular recesses

dizziness, fatigue, palpitation or sudden cardiac arrest. LVNC with coexisted cardiac anomaly, such as myocardial bridge, coronary-pulmonary fistula, single coronary artery disease or significant atherosclerotic coronary arteries is more rarer than isolated LVNC (Zhu et al., 2018; Sun et al., 2019). However, ALCAPA concomitant with LVNC is not reported in the literature to our knowledge. Here we report a survived case of ALCAPA concomitant with LVNC in a young half-marathon runner.

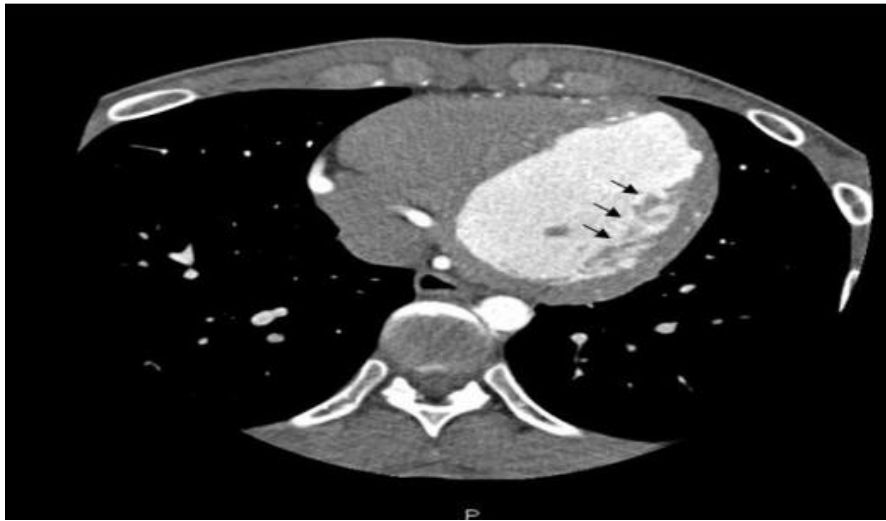
### CASE REPORT

We report the case of a 22-year-old man half marathon runner, who had no history of systemic disease, such as hypertension, diabetes or chronic airway disease in the past. This young man is a half-marathon player and his

best record in half-marathon competition is about 100 minutes. He came to our hospital for health examination of enlist into the army. In the regular medical check-up, he presented stable vital signs: a blood pressure of 116/71 mmHg, heart rate of 59 bpm, respiratory rate of 12 bpm, Glasgow Coma Scale Score of 15 and Body Mass Index (BMI) of 19.3(kilogram/meter<sup>2</sup>). No significant abnormal finding in general appearance, chest X-ray and blood sample. Electrocardiography revealed normal sinus rhythm with signs of high lateral (Q wave negative T wave in aVL and RS aspect in V1 lead) and apical (Q wave in V3 and V4 leads) previous myocardial infarction (Figure 1). Echocardiography demonstrated 81% of ejection fraction (EF), minimal mitral valve prolapse with trivial mitral valve regurgitation and multiple prominent ventricular trabeculations with deep intertrabecular recesses ( Figure 2). Thallium-201 myocardial perfusion single-photon



**Figure 3:** Left coronary artery origin from pulmonary artery. Ao: aorta, PA: pulmonary artery, LAD: left anterior descending coronary artery, LCX: left circumflex coronary artery, RCA: right coronary artery



**Figure 4:** Multiple trabeculatiois (arrow) within left ventricle.

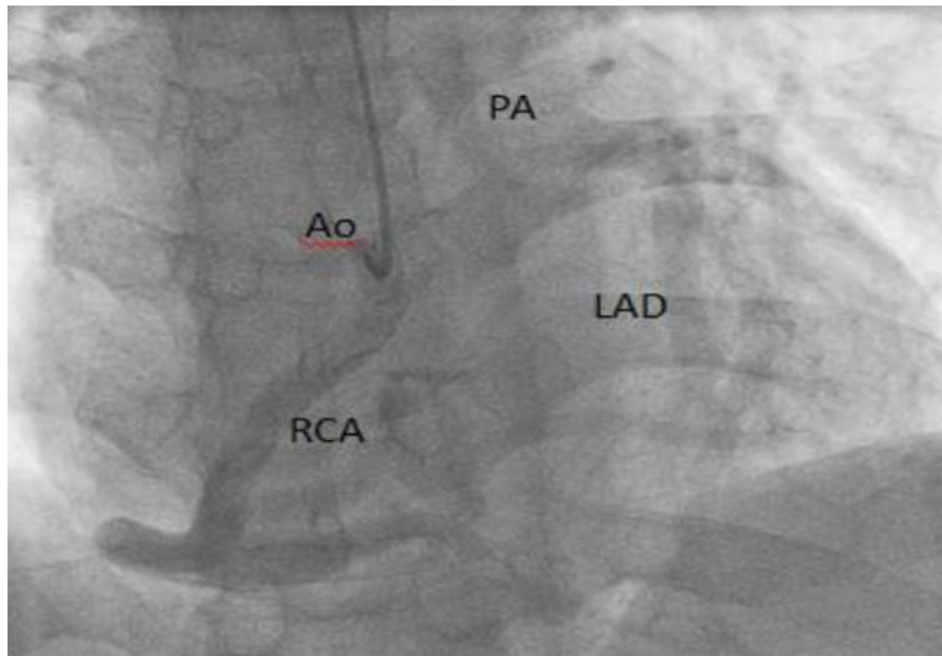
emission computed tomography (SPECT) displayed decreasing myocardium perfusion in extensive anterior wall, apex, and inferior wall of the left ventricle. Coronary computed tomography angiogram (CTA) revealed left coronary artery origin from pulmonary artery with compensatory enlarged right coronary artery and tortuous inter-coronary collateral vessels (Figure 3). Multiple trabeculations with deep recesses of left ventricle (Figure 4) was also detected via coronary CTA image. Coronary artery angiography (CAG) also showed the similar anatomy abnormality (Figure 5) as coronary CTA images and had no meaningful stenotic coronary artery manifestation. Due to the above abnormal finding, preventive bypass surgery was indicated for this half marathon runner.

However, the patient indeed refused to receive heart operation after full discussion. At follow-up one year, he was doing well under proper life modification after diagnosis and then he kept training himself for sports

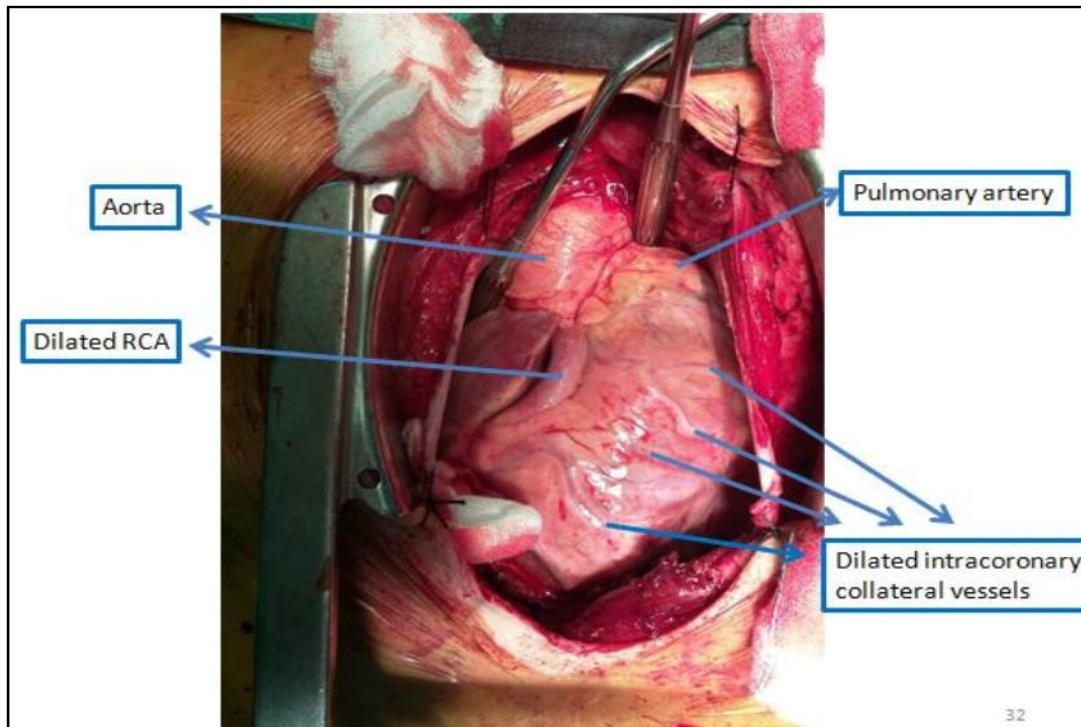
activities as before. Unfortunately, the patient suffered an out-hospital cardiac arrest (OHCA) during a half-marathon competition two years later. The patient survived after advanced cardiovascular life support (ACLS) management and he received emergent coronary artery bypass graft (CABG) surgery then. Cardiac vessel anomaly was found as similar as coronary CTA and CAG during surgery procedure (Figure 6). Ligation of left main coronary artery (LMCA) orifice in pulmonary artery and bridging vessel graft from aorta root to left anterior descending artery (LAD) were performed successfully. He was discharged 18 days after hospitalization with preserved left ventricular function. At follow-up three years, he was doing well under optimization medication.

## DISCUSSION

ALCAPA was first reported in 1886 by Brooks, and its first



**Figure 5:** Enlarged right coronary artery and left coronary artery origin from pulmonary artery. Ao: aorta, PA: pulmonary artery, LAD: left anterior descending coronary artery, RCA: right coronary artery.



**Figure 6:** Left coronary artery origin from pulmonary artery with compensatory dilated right coronary artery and dilated collateral vessels

clinical description associated with the results of the autopsy was described by Bland et al. in 1933. ALCAPA is an extremely rare coronary artery malformation in the world. ALCAPA often can cause potential myocardial

ischemia, subsequently leading to heart failure, mitral valve insufficiency or arrhythmia. Connexin 43 alpha-1, a member of the family of the gap junctions, has an important role in the development of the coronary artery



in mice. Sawaya et al. proposed that the mutation of the 6q22-31 gene encoding this protein may have a role in ALCAPA in humans, but none of their four cases showed any abnormality in this gene.

They call for more investigations in this field. However, spongy myocardium as we known LVNC was first declared during an autopsy in 1932 by Bellet et al. And the first case demonstrated in a living patient was reported by (Engberding et al., 1984). LVNC is indicated an embryo arrest phenomenon (Jenni et al., 1986). Three mainly clinical manifestations are heart failure, arrhythmia and embolic events. Genetic defect is believed to be a possible mechanism of LVNC, possible genetic loci involved the G4.5 gene of the Xq28 chromosome, the alpha-dystrobrevin gene, cardiac-specific gene CSX, mutated chromosome 10q22, distal chromosome 5q deletion or the FKBP12 gene (Brian et al., 2004). However, almost 10% of athlete full-fit the criteria of echocardiographic LVNC in the past research by Gati et al. (2013). Pressure overloading phenomenon of cardiac and genetic variation may play possible mechanisms in increased LV trabeculation formation, especial in black athletes.

To our knowledge, this patient is the first case who had ALCAPA concomitant with LVNC in the world. The patient may have some one or more mutated share-gene locus theoretically. So, the patient could check gene geography, family members can also do. Therefore, according to the prior studies, the objective of bypass surgery is suitable to restore coronary artery circulation and myocardial perfusion. Coronary transfer proposed by Kai Ma et al. in 2014 is also an alternative choice for these patients. Therefore, the therapy of heart failure combination is extremely important even during controlled disease.

However, a lot of young men will serve in the army of the worldwide yearly. It is very important to arrange exhaustive medical examination, such as physical examination, ECG, chest X-ray and blood sampling to early detect possible fatal diseases before military service. Many Teens-diseases and congenital diseases may be found out underwent health check-up, such as diabetes, thalassemia, asthma, Wolff-Parkinson-White syndrome (WPW syndrome), LVNC and ALCAPA, like our case. The higher socioeconomic impact of pre-military health check-up will achieve.

In our case, the dilemma of a choice between preventive cardiac surgery and conservative therapy in ALCAPA is a true embarrassment. According to the prior studies, the objective of bypass surgery is suitable to restore coronary artery circulation and myocardial perfusion Boutsikou et al. (2018); Alasrawi (2018); Qiu et al. (2016). Therefore, the therapy of heart failure combination with optimal medication and preventive intracardiac defibrillator (ICD) or cardiac resynchronization therapy (CRT) implantation is also a puzzle for patients of LVNC Kimura et al. 2018), especial in those asymptomatic ones. To my knowledge, ICD implantation may be an acceptable clinical advice initially for lethal dysrhythmia therapy in asymptomatic patients, like our case.

In conclusion, early detection of this clinical entity via screening health examination and intensive management of symptomatic patients with cardiac surgery may result in a favorable outcome. Besides, family-genetic geography testing may make sense in this kind of genetic disorders.

### Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of the paper.

### REFERENCES

- Alsara O, Kalavakunta JK, Hajjar V, Alsarah A, Cho N, Dhar G (2014). Surviving sudden cardiac death secondary to anomalous left coronary artery from the pulmonary artery: A case report and literature review. *Heart & Lung* 43: 476-480
- Boutsikou M, Shore D, Li Wu, Rubens M, Pijuan A.(2018).Anomalous left coronary artery from the pulmonary artery (ALCAPA) diagnosed in adulthood: Varied clinical presentation, therapeutic approach and outcome.*International J. Cardiol.* 261(15):49-53.
- Brian C, Weiford, Vijay D, Subbarao, Kevin M, Mulhern, (2004). Noncompaction of the Ventricular Myocardium *Circulation.* 109:2965-2971.
- Gati S, Chandra N, RL Bennett RL, Reed M, Kervio G, Panoulas VF, Ghani S, Sheikh N, Zaidi A, Wilson M, Papadakis M, Carré F, Sharma S (2013). Increased left ventricular trabeculation in highly trained athletes: do we need more stringent criteria for the diagnosis of left ventricular non-compaction in athletes. *Heart* 99:401-408.
- Jenni R, Goebel N, Tartini R, Schneider J, Arbenz U, Oelz O (1986). Persisting myocardial sinusoids of both ventricles as an isolated anomaly: echocardiographic, angiographic, and pathologic anatomical findings. *Cardio Vascular and Interventional Radiology,* 9(3):127-131.
- Kimura M, Kawano K, Yaoita H, Kure S (2018). Successful Treatment of an Infant with Left Ventricular Noncompaction Presenting with Fatal Ventricular Arrhythmia Treated with Cardiac Resynchronization Therapy and an Implantable Cardioverter Defibrillator. *Case Reports in Cardiology* Volume 2018, Article ID 9562326.
- Sun L, Zhao E, Wei Y, Kang C, Liu B (2019). Thickness and Ratio of Noncompacted and Compacted Layers of the Left Ventricular Myocardium Evaluated in 56 Normal Fetuses by Two-Dimensional Echocardiography. *BioMed Research International* Volume 2019, Article ID 3726846.
- Liu Y, Chen H, Shou W (2018). Potential Common Pathogenic Pathways for the Left Ventricular Noncompaction Cardiomyopathy (LVNC). *Pediatric Cardiology* 39:1099-1106.
- Qiu J, Li S, Yan J, Wang Q, Song Y, Sun H, Li D (2016). Repair of anomalous coronary artery from the

- pulmonary artery: A-signal center 20-year experience. *International J. Cardiol.* 223: 625–629.
- Samah A (2018). Good Prognosis of ALCAPA (Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery Syndrome) with early Diagnosis and Surgical Treatment. *Global Journal of Medical Research* 18(3):1.
- Xiangming Z, Yang Y, Guobing H (2018). Left ventricular noncompaction in patients with coronary artery disease: Preliminary analysis of echocardiographic findings. *J Clin. Ultrasound* 46:475-479.
- Yamanaka O, Hobbs RE (1990). Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn.* 21(28):e40.