The Survival of an Old Lady with ALCAPA

Chaothawee L, MD
email: augustl2509@gmail.com

Lertlak Chaothawee, MD[1]
Gumpanart Veerakul, MD[2]
Manasawee Indrabhinduwat, MD[2]

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[1] Cardiac Imaging Unit, Cardiology department, Bangkok Heart Hospital, Bangkok Hospital Group, Bangkok, Thailand
[2] Cardiovascular Research and Prevention Center, Bhumibol Adulyadej Hospital RTAF, Bangkok, Thailand

**Address Correspondence to author:*
Cardiac Imaging Unit, Bangkok Heart Hospital,
2 Soi Soonvijai 7, New Petchburi Road, Bangkapi, Huaykwang,
Bangkok 10310, Thailand.
E-mail: augustl2509@gmail.com

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The coronary artery system is composed of two major systems, the left and right coronary artery system. Both left and right coronary arteries originate from the aortic left and right coronary cusp respectively. The left anterior descending artery (LAD), the main vessel of the left coronary artery system, is considered to be the most important of the coronary artery system. The LAD supplies the left ventricular anterior wall that plays the main role in the left ventricular pumping function. The right coronary artery system is also considered to be the second most important system that mainly supplies the left ventricular inferior wall. The balance of myocardium blood supply demands an adequate coronary artery blood supply from both systems. If the blood supply to some portion of the myocardium is entirely blocked or the energy demand is much higher than the supply, severe myocardium injury or sudden cardiac death may occur. Total blocking of the left main coronary artery (LMA) and proximal LAD vessel is the main cause of death. **How does a person survive with a total absence of arterial blood supply from the left coronary artery system?**

ALCAPA syndrome is an example of a congenital abnormal condition where a person stays alive even in the absence of blood supply from the left coronary artery system. ALCAPA stands for "**A**nomalous origin of the **L**eft **C**oronary **A**rtery from the **P**ulmonary **A**rtery".[1] ALCAPA is also called **Bland-White-Garland syndrome** because it was described by Bland and colleagues in 1933 during an autopsy.[1] By definition, the main key abnormality of the ALCAPA syndrome is the origination of the left coronary artery system coming from the main pulmonary artery instead of the aortic coronary sinus. ALCAPA is a rare condition and the incidence accounts for 0.25-0.5% of all congenital heart disease cases.[2] This abnormal condition develops during the period of embryogenesis and it may be due to the abnormal septation of the conotruncus into the aorta and pulmonary artery or the persistence of the pulmonary buds together with involution of the aortic buds that form the coronary arteries.[2] The left coronary system is retrogradely supplied by the collaterals from the right to left. ALCAPA syndrome results in coronary artery steal phenomena and a left to right blood shunting which occurs after birth. Coronary steal phenomena and left to right blood shunting are caused by the entering of the blood in the LAD lumen (artery) into the low resistant pulmonary artery trunk (venous). The pulmonary artery steals the blood from the heart, hence, the left ventricular myocardium is underperfused.[3,4] ALCAPA syndrome has two types, the infant type and adult type. In the infant type, about 90% die within the first year of life due to myocardial infarction and heart failure. The development of a collateral system between the RCA to LAD also appears after birth and it is the **key to life**, if a collateral system does not develop well enough on time, 90% of patients die during

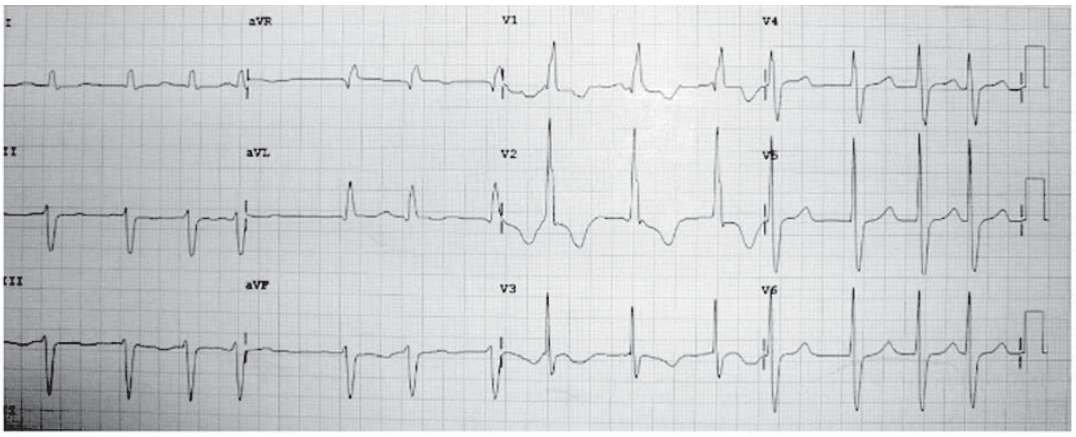

Figure 1: *ECG on admission showed atrial fibrillation, RBBB with secondary ST-T changes.*

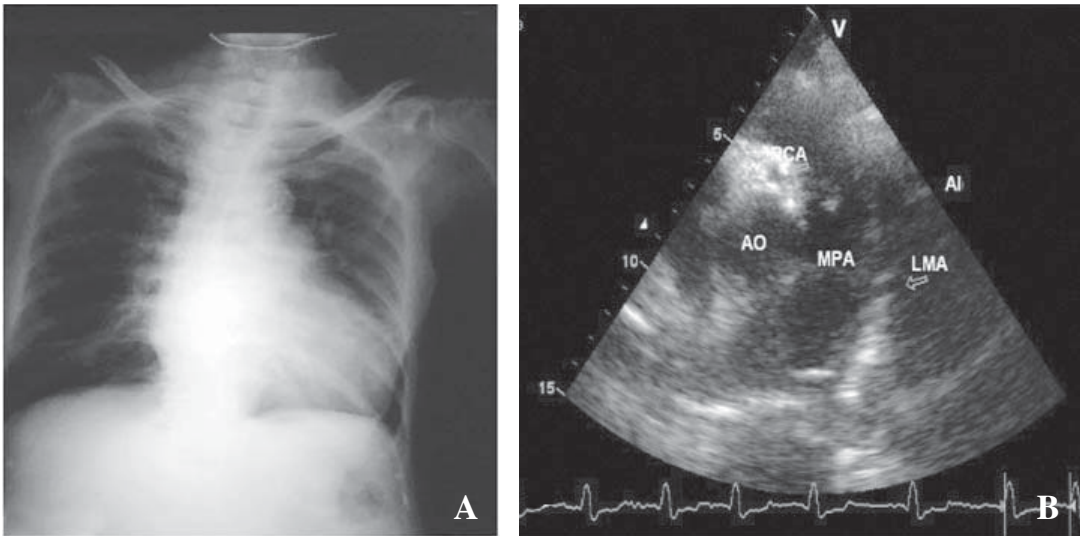

Figure 2: *Left: Chest film five months before this event showed cardiomegaly and prominent pulmonary trunk (A). Right: short axis view of the aortic root and pulmonary trunk that showed origin of RCA and LMA.*

infancy shortly after birth from congestive heart failure and myocardial infarction[4] because the collateral system is the only transport pathway to bring arterial blood from the right coronary artery to the left coronary artery. The ALCAPA patient who outgrows death in the young, may be asymptomatic or present with myocardial ischemia, heart failure, chronic mitral regurgitation and sudden death.[5] Although most ALCAPA patients die young, the survival rate and prognosis can be improved by early detection using noninvasive methods such as echocardiography.[6] Surgical correction should be considered soon after verification of the diagnosis.[5]

Case Report

A 79-year-old Thai lady, had a known history of diabetes mellitus, hypothyroidism, chronic atrial fibrillation and suffered from right sided ischemic stroke for over ten years, presented in May 2011 with pulmonary edema. Physical findings of the left heart failure correlated well with the chest film. Pan systolic murmur grade 2/6 was audible at the apical area. ECG showed atrial fibrillation with moderate ventricular rate response, complete right bundle branch block (RBBB) and precordial ST-T segment depression, (Figure 1). Echocardiography revealed an enlarged left ventricle, dilated left atrium, anterior mitral valve prolapsed causing moderate to severe mitral regurgitation, globally decreased LV systolic function, EF of 0.44 with antero-apical hypokinesia and elevated pulmonary artery pressure, 60 mmHg. With medication, she was successfully extubated, transferred to an intermediate unit for a few days and had recurrent pulmonary congestion requiring re-intubation. Similar events repeatedly occurred three times so it was decided to exclude coronary stenosis by conventional angiography (CAG). CAG images (Figure 2A-B) revealed no left main artery originated from the aortic root. A huge single right coronary artery supplied the whole ventricle and transeptally filled the left coronary (LCA). It was suspicious that the left main (LM) might originate from

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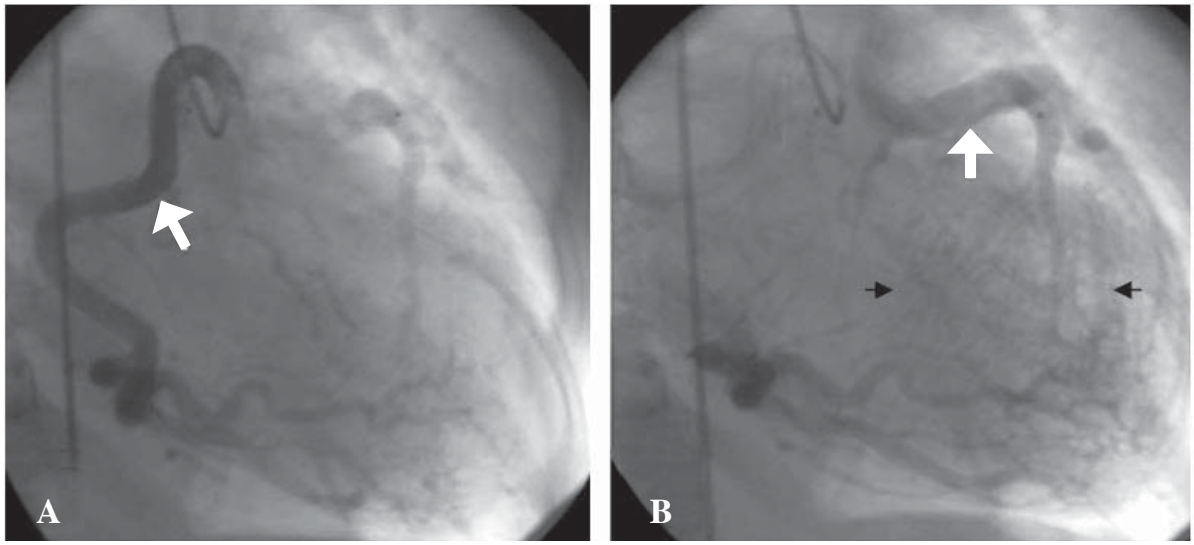

Figure 3: *Left heart CAG images revealed (A) the RCA which was in the right position (A) with no show of the left coronary artery origin at the left cusp sinus , (B) the left coronary arteries (white arrow) were supplied by the right to left collaterals (black arrow).*

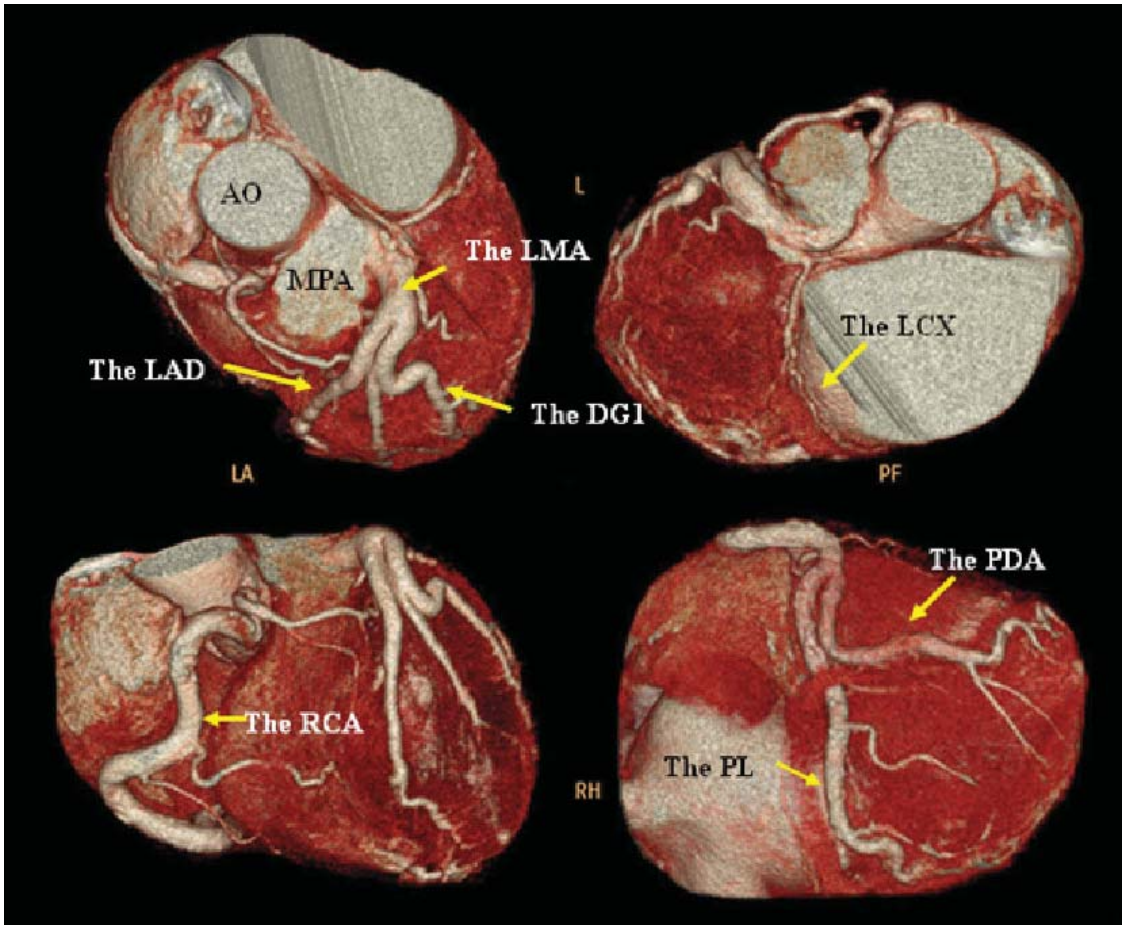

Figure 4: *Volume rendering images of the 256- slice MDCT revealed the large RCA which originated from the right cusp and the LMA originated from the MPA (ALCAPA).*

AO= Aorta *MPA= Main pulmonary artery* *LMA = Left main artery*
LAD = Left descending coronary artery *LCX= Left circumflex coronary artery*
RCA = Right coronary artery *PL= Posterior lateral artery*
PDA = Posterior descending artery

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the main pulmonary artery so she was referred for a multi-detector computerized tomography (MDCT) study. A 256 detector CT scanning with iodinated contrast injection was requested to define the diagnosis and it showed an enlarged RCA took off from the right aortic cusp as shown in Figure 3. The RCA gave the long postero-lateral (PL) and the posterior descending (PDA) branches, supplying the lateral and the posterior wall of the left ventricle, retrogradely filled the left anterior descending (LAD) and the LM arteries. Contrast phase image of MDCT scan confirmed the abnormal origin of the LM trunk that originated from the main pulmonary artery and supplied the small left circumflex (LCx) artery. The course of the LAD and the LCx arteries were still running along the inter-ventricular and the atrio-ventricular groove respectively. Based upon the above MDCT findings, the final diagnosis was ALCAPA syndrome. In ALCAPA cases, besides a prominent pulmonary trunk in the chest film, an echocardiogram may provide some diagnostic clue. In the short axis view of the aortic root, there was no left main origin (see Figure 2B). By color Doppler echocardiography, the pulmonary artery shows the retrograde flow in the pulmonary artery and systolic-diastolic flow through the septal collaterals which indicated that the blood supply to the left main artery and the LAD was through the pulmonary artery (Figure 3). The ECG gated MDCT offers a direct visualization of both the origin of the RCA and LCA, and provides a definitive diagnosis (Figure 4). In our case, the flourish right to left collateral supply noted in conventional angiogram and MDCT images explained why she had been living well through to her 8th decade. The significant mitral regurgitation, either resulted from anterior mitral valve prolapse, combining with dilated ventricle secondary from ischemic cardiomyopathy or both, created a high LVEDP and LA pressure and contributed to recurrent CHF. In addition, the high PAP, 60 mmHg, further compromised myocardial perfusion since the LCA received the reverse flow from RCA. After an uneventful tracheostomy, she was clinically stable and had avoided heart failure for months. The family decided to treat her conservatively.

Discussion

The diagnosis was easily overlooked by routine and non-invasive examinations. The echocardiograms might not have provided a definite diagnostic. The coronary angiogram provided a clue but does not always show a precise abnormal origin of the LM artery. MDCT can be used as a first diagnostic or a complimentary noninvasive technique since it displays the origin of both arteries, the course and other associate diseases i.e. patent ductus arteriosus (PDA), tetralogy of fallot (TOF), ventricular septal defect (VSD) and atrial septal defect (ASD). In a low calcium scoring patient, MDCT can be a one-stop tool for a definitive diagnosis before surgical planning.

Conclusion

Although ALCAPA syndrome is a very rare condition it can be accidentally found in everyday practice. This article illustrated a surviving case in her 8th decade before succumbing to heart failure from multiple causes. It is very rare for ALCAPA patients to survive to their sixth or even seventh decades of life.[7] The key to the survival of this patient is the well developed collaterals from the right to left. Awareness of this condition may prevent complications and may improve survival rates and prognosis.

References

1. Bland EF. Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. *AM Heart J* 1933;8:787-801.
2. Dodge-Khatami A, Mavroudis C, Backer CL. Anomalous origin of the left coronary artery from the pulmonary artery: collective review from surgical therapy. *Ann Thorac Surg* 2002;74:946-55.
3. Lin C-P, Chen Y-P, Chen T-H, et al. Anomalous origin of the left main coronary artery from the main pulmonary artery in a young adult. *Circulation* 2001;104:1575-6.
4. Wesselhoeft H, Fawcett JS, Johnson AL, et al. Anomalous origin of the left coronary artery from the pulmonary trunk: its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. *Circulation* 1968;38:403-5.
5. Tang WC,Chung YC, Sung-How S, et al. Adult type anomalous origin of the left coronary artery from the pulmonary artery. *Acta Cardiol Sin* 2007;23:115-8.
6. Fontana RS, Edwards JE. Congenital Cardiac Disease: a Review of 357 Case Studies Pathologically. *WB Saunders* 1962:291.
7. Purut CM, Sabiston DC Jr. Origin of the left coronary artery from the pulmonary artery in older adults. *J Thorac Cardiovasc Surg* 1991;102:566.

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