· CASE ANALYSIS · · 临床病例讨论 ·

DOI:10.11817/j.issn.1672-7347.2017.12.016
www.csumed.org/xbwk/fileup/PDF/2017121458.pdf

Successful treatment in a patient with sudden cardiac arrest due to anomalous left coronary artery arising from pulmonary artery

ZHOU Liping, LI Xiaogang, QI Min, WANG Aimin

(Emergency Department, Xiangya Hospital, Central South University, Changsha 410008, China)

ABSTRACT To enhance the understanding of the left coronary artery originating from the pulmonary artery, we report a case of sudden cardiac arrest during exercise. After successful cardiopulmonary resuscitation, the patient underwent echocardiography and selective coronary angiography, and the disease was firmly diagnosed. Consequently, a thoracic surgery was performed. During the operation, the left coronary artery was transplanted to the root of the aorta and the pulmonary valve and artery were reconstructed. Finally, the surgery was successful.

KEY WORDS anomalous left coronary artery from pulmonary artery; coronary angiography; congenital heart disease

左侧冠状动脉起源于肺动脉患者心脏骤停成功救治1例

周利平，李小刚，祁敏，王爱民

(中南大学湘雅医院急诊科，长沙 410008)

[摘要] 为增强对左侧冠状动脉起源于肺动脉的认识，报告1例该病的病例资料。该例成年患者在运动中发生心脏骤停，心肺脑复苏成功后行心电图、心脏彩超、心脏冠状动脉计算机体层摄影及选择性冠状动脉造影检查后证实为左侧冠状动脉起源于肺动脉。该患者被转至胸外科行外科手术，术中术者直接将异常起源肺动脉的左冠状动脉移植到主动脉根部上，并重建肺动脉瓣窦和肺动脉。手术治疗取得成功。

[关键词] 冠状动脉异常起源于肺动脉；冠状动脉造影术；先天性心脏病

An anomalous left coronary artery originating from the pulmonary artery (ALCAPA), first discovered in 1911, is a rare congenital malformation. Because of the physiological characteristics of this disease, approximately

Date of reception: 2016-09-04
First author: ZHOU Liping, Email: 2661094304@qq.com
Corresponding author: LI Xiaogang, Email: lixiaogang@medmail.com.cn

95% of the patients develop congestive heart failure and die in infancy. Individuals who survive into childhood or adulthood may experience sudden cardiac arrest after exercise. In the past years in China, the prognosis for these patients was poor because of insufficient knowledge and experience in treating this disease. In recent years, with the rapid improvement in the diagnosis and treatment of congenital heart diseases, the success rate of treatment for ALCAPA has significantly improved[1].

1 Case presentation

A 21-year-old male patient suddenly fainted during long-distance running and remained unconscious when participating in an organized activity. He was sent to the hospital a few minutes later, and after examination, the diagnosis was cardiopulmonary arrest. Cardiopulmonary resuscitation was immediately performed, and approximately 5 minutes later, the patient's breathing and heartbeat were re-established. After successful resuscitation, the patient was in a mild coma, with impaired limb movement. A physical examination showed a pulse rate of 94 beats per minute, a respiratory rate of 20 beats per minute, and a blood pressure of 120/80 mmHg (1 mmHg=0.133 kPa). The patient appeared to recover normally, except for a state of mild coma, and was uncooperative during the examination. The trachea was centrally located, and the thorax was symmetrical with no deformity. The patient had coarse breathing sounds in both lungs, moist rales in the lung bases, and increased sputum rales in the throat. No precordial bulging was observed. The apex beat was localized in the fifth left intercostal space in the mid-clavicle line. Heart rate was 94 beats per minute. The heart rhythm was normal, and a Grade 2/6 "blowing" systolic murmur was heard at the cardiac apex and pulmonary valves. Limb movement was impaired with Grade 1 muscle strength and increased muscle tone. The patellar reflex was intact, whereas the Kernig, Babinski, and Brudzinski signs were negative. The diagnostic considerations included hypoxic-ischemic encephalopathy after cardiopulmonary resuscitation and pulmonary infection.

Second phase treatment included cerebral resuscitation, early hyperbaric oxygen treatment after patient's vital signs were stabilized, dehydration to reduce brain cell swelling, hypothermia for neurologic protection, edaravone to scavenge oxygen free radicals, and a xingnaojing injection to promote brain cell metabolism. Neural function recovered, and the patient began to answer simple questions 10 days after the treatment. However, his articulation was unclear. He regained voluntary movement of his legs and left upper arm. The right upper arm showed higher muscle tone with limited movement. Electrocardiography and cardiac color Doppler ultrasound were performed during the treatment.

An abdominal B-mode ultrasound showed no obvious abnormalities in the liver, gallbladder, spleen, pancreas, kidneys, or ureters. The result of cranial computed tomography (CT) scan was normal.

A routine blood test showed a white blood cell count of 11.7×10^9/L and a neutrophil count of 8.8×10^9/L (75.4%). Routine urinalysis indicated urine urobilinogen of 3+, protein +, and vitamin C level of 2+. Liver and kidney function indices and myocardial enzymes were as follows: total bilirubin, 23.3 μmol/L; direct bilirubin, 10.2 μmol/L; alanine aminotransferase, 55.1 U/L; aspartate aminotransferase, 62.2 U/L; lactate dehydrogenase, 427.4 U/L; creatine kinase, 1 025.2 U/L; creatine kinase isoenzyme, 21.4 U/L; and myoglobin, 165.6 μg/L.

An anteroposterior chest radiograph showed lung exudative lesions. Color Doppler echocardiography showed right ventricle, 17 mm; left ventricle, 64 mm; left atrium, 30 mm; aorta, 30 mm; pulmonary artery, 21 mm; right atrium, 39 mm×37 mm; interventricular septum (IVS), 11 mm; left ventricular end diastolic posterior wall, 7 mm; and inferior vena cava, 16 mm. Heart measurements were as follows: Stroke volume, 83 mL; cardiac output, 7 L/min; ejection fraction (EF), 45%; and fractional shortening, 23%. Two-dimensional (2-D) ultrasound examination showed increased left ventricular diameter, and the other chambers were of normal size. Diameters of the main aorta and pulmonary artery were normal. Valve morphology in both the open and closed positions was normal. The interventricular and interatrial septal echoes were continuous. The thicknesses of the IVS and left ventricular posterior wall were normal, with reversed movement and a slightly reduced range of motion. In the resting state, no obvious segmental motion abnormality of the left ventricle wall was observed. The left ventricular long-axis view showed that the diameter of the initial right coronary artery segment was widened, with a diameter of approximately 8.5 mm, and was not clear inside. The short-

axis view showed that the diameter of the left coronary artery was approximately 6 mm. No obvious abnormalities were observed in the aortic arch or descending aorta. Color Doppler echocardiogram showed a large amount of blood flow in the left ventricular myocardium; a blue jet of retrograde blood flow (mild) at the mitral and tricuspid valves during systole, and a red jet of retrograde blood flow (mild) at the aortic valves during diastole. The examinations indicated left ventricle enlargement, mild mitral, tricuspid, and aortic valve regurgitation, reduced left ventricular function (EF 45%), and widened right coronary artery with the possibility of leakage in small coronary branches (Figure 1).

Electrocardiogram (ECG) showed sinus tachycardia, left deviation of elective axis, high voltage of left ventricular surface (Figure 2).

A Holter monitor test showed 24 occasional twice in pairs, one run of tachycardia contractions including

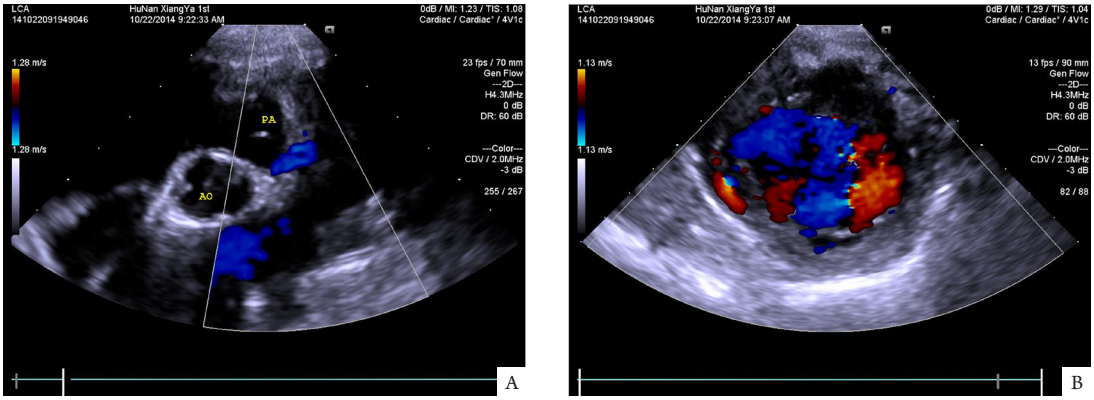

Figure 1 Color Doppler echocardiography

A: Echocardiogram shows the left coronary artery originaing in the pulmonary artery, in which the blood flow is reversed perfusion. B: Left ventricular short axis section shows abundant collateral coronary flow signals

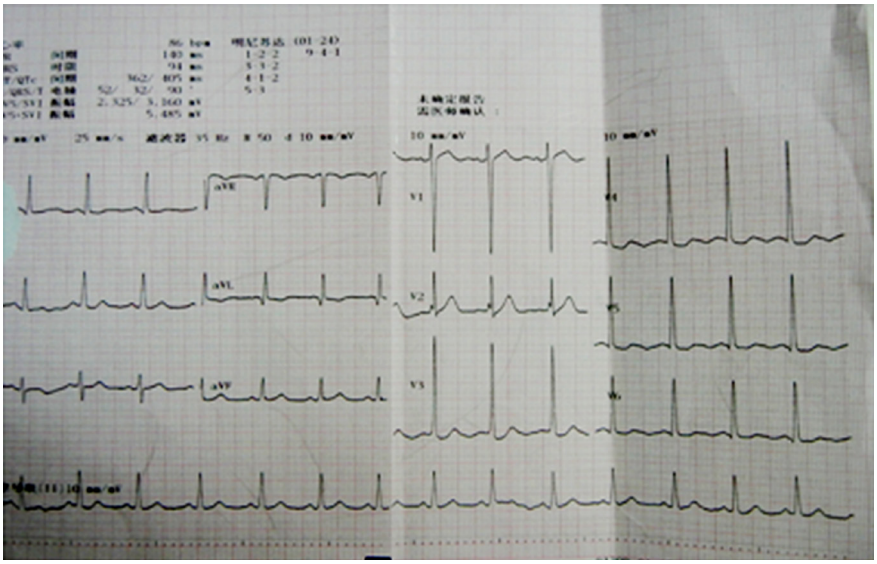

Figure 2 Electrocardiogram of the patient

twice in pairs, one run of tachycardia, no bi- or trigeminy, and occasional ventricular premature contractions, 1 mm persistent horizontal ST segment depression in the V5 lead, persistent flat T-waves, and decreased heart rate variability.

A brain MRI scan showed that compaed with the previous scan, the patchy abnormal signals in both basal ganglia had disappeared, the bilateral gyrus swelling had significantly improved, the morphology and size of the arachnoid cyst at the left temporal pole were basically unchanged, the bilateral sinus inflammation had improved, and the bilateral mastoiditis was slightly worse. Other conditions observed were unchanged.

The anteroposterior chest radiographs showed increased and abnormal lung markings, but no obvious pathological changes were observed. The heart and diaphragm were normal.

The results of coronary angiography were as follows: Coronary artery distribution with right coronary artery dominance; the left coronary artery opening in the pulmonary artery, with a diameter of the proximal segment of 5.5 mm; the right coronary artery with full expansion, with the proximal segment diameter of 12 mm, middle segment diameter of 9 mm, and distal segment diameter of 7 mm; and a blood circulation pattern of aorta→right coronaryartery→left coronary artery→pulmonary artery. The right ventricular angiogram showed enlargement of right ventricle and left atrium and expansion of pulmonary artery. Conclusions based on the angiograms were right coronary artery expansion, a left coronary artery originating from the pulmonary artery, right ventricle and left atrium enlargement, and pulmonary artery expansion.

The results of electroencephalography performed on January 3, 2012, were generally abnormal.

Antibiotics, brain protection, and enteral nutrition support therapies were given. A hyperbaric oxygen treatment was administered once daily. The patient recovered consciousness. However, the intellectual level declined and enunciation was unclear. The patient could feed himself and get out of bed and walk when assisted. Further treatment in preparation for heart surgery included continued hyperbaric oxygen therapy, rehabilitation therapies to improve limb control, and therapy to improve mental acuity.

After successful cerebral resuscitation, the patient was mentally normal with mobility in all 4 limbs and basic self-care abilities, but the intellectual level declined. His vital signs were stable, and he was transferred to the thoracic surgery department. Preoperative computed tomography angiography image scan was abnormal. The image showed right coronary artery expansion, a left coronary artery originating from the pulmonary artery, right ventricle and left atrium enlargement, and pulmonary artery expansion (Figures 3). During surgery, the anomalous left coronary artery from the pulmonary artery was grafted directly to theaortic root, and the pulmonary valve sinus and pulmonary artery were reconstructed. The left coronary blood flow was restored (Figure 4).

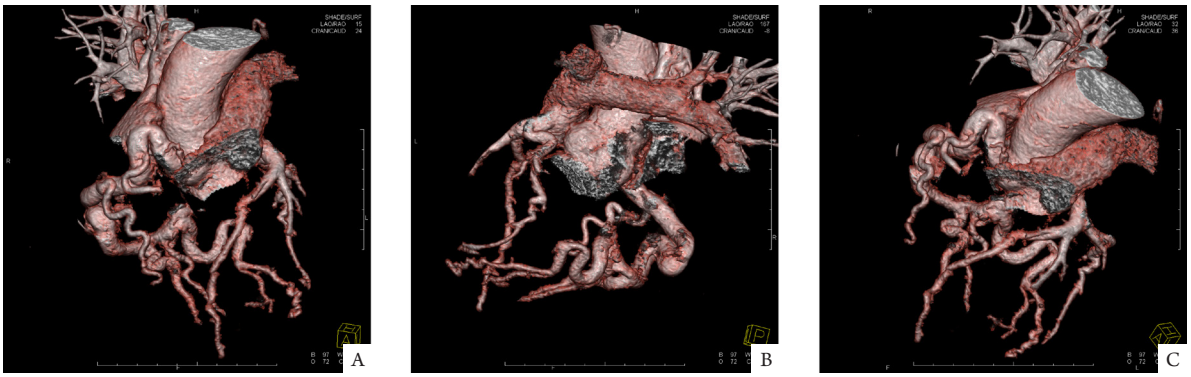

Figure 3 Preoperative heart computed tomography angiogram

A: Right coronary artery expansion; B: A left coronary artery originating from the pulmonary artery; C: Pulmonary artery expansion

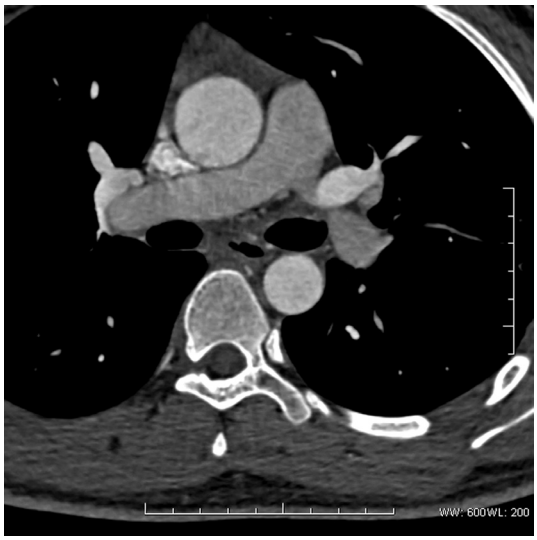

Figure 4 Three-dimensional images of the aorta after reconstruction surgery

2 Discussion

An ALCAPA is a rare congenital cardiac malformation with incidence rate of 1/30 000 to 1/300 000[2-3] and accounts for 0.5% of the congenital heart diseases. The prognosis of this disease is very poor, and 90% of the patients die within 1 year old[4], thus early diagnosis is extremely important. If the patient survives to adulthood, left ventricular failure or sudden cardiac arrest are likely to occur. A consensus has not been reached for surgical approaches to this disease, which are mainly divided into simple ligation of the proximal origin of the anomalous coronary artery and coronary revascularization. This operation is simple, and can prevent "coronary stealing" of the left coronary blood flow and improve myocardial perfusion. However, when coronary atherosclerotic lesions affect the right coronary artery in old age, serious complications may occur. In addition, this treatment is unsuitable for infants with no obvious left to right coronary artery shunt. Therefore, at present, most surgeons feel that two coronary systems should be established if possible.

Clinically, the patient's location of the opening of the left coronary artery which originating from the pulmonary root was abnormal. However, the distal branch morphology was normal. The right coronary artery was normal, with the opening located in the ascending aorta, and had normal branching and a rich collateral circulation. Blood flow originated from the opening of the right coronary artery→extensive coronary collateral circulation→left coronary artery→pulmonary artery. Although the blood oxygenation level in the left coronary artery was good, left ventricular ischemia was apparent. The "stealing" of blood flow from the left ventricle was the result of the collateral circulation flowing into the low-pressure pulmonary artery. Multiple foci of left ventricular necrosis were observed, with a transition from old to new, accompanied by extensive sub-endocardial fibrosis and calcification, which often affects papillary muscles resulting in mitralvalve regurgitation. Mitral valve regurgitation could also be caused by mitral annulus widening. If collateral circulation between left and right coronary arteries is large, patients may survive to childhood or even longer, but may exhibit a right coronary artery of normal origin with branches significantly enlarged and tortuous, sometimes manifested as cirsoidangioma.

Regarding clinical symptoms of this disease, approximately 80% of patients suffer heart failure in infancy. Patients who survive after 1 year into childhood and adulthood show various degrees of symptoms, with some patients being asymptomatic during intense activities. The majority of patients experienced chest tightness upon exertion and shortness of breath, sometimes accompanied by angina pectoris; although cases of congestive heart failure are rare, except the patient with significant mitral regurgitation. Clinical signs include heart failure and mitral valve regurgitation in infancy, mainly after 1 year old, consisting of a systolic murmur or double murmur in the pulmonary valve auscultation area, with tremors in some patients, which results from a coronary artery right to left shunt to the pulmonary artery. Some patients have a mitral valve regurgitation murmur.

Supplementary examinations for patients with an ALCAPA are important for diagnosis. Among them, aortic root angiography is the most important test[3].

The rationale for performing the angiographic assessment is to obtain overview of the left and right coronary arteries, collateral circulation, and the location of anomalous coronary arteriesin the main pulmonary trunk. Contrast agents that enter into the right coronary artery fill the left coronary artery through retrograde flow between coronary arteries and then enter the low-pressure pulmonary artery. The right coronary artery is clearly

dilated. Unless the patient exhibits severe pulmonary hypertension, pulmonary angiography cannot be used for imaging coronary arteries with anomalous origins. When one coronary artery solely originates from the pulmonary artery, diagnosis based on angiography of the aortic rootis not difficult. However, anomalous origins of coronary artery branches from the pulmonary artery can be easily overlooked in theaortic root angiogram[4-5]. Therefore, selective coronary angiography needs to be performed when necessary that can reveal the trunk, its branches, and their connections to the pulmonary artery in detail.

ECG basically reflects ischemic changes in the area of the anomalous coronary artery. Some patients may not show ischemic changes in the ECG. However, when there are ischemic changes they should be identified.

Echocardiography can provide important clues to disease diagnosis. Diagnostic accuracy depends on skills of operators and their pathological and hemodynamic knowledge about the disease. The characteristic changes are pulmonary artery exploration, abnormal blood flow, and abnormal blood vessel connections. Because the pressure has been reduced by vascular graft anastomoses, shunting decreases, the blood flow velocity in the abnormal connections decreases, generally not exceeding 1.5 m/s. In addition, the anomalous coronary artery generally originates at the pulmonary artery root, which is different from patent ductus arteriosus. A single, dilated, tortuous coronary artery is an indirect characteristic of the disease.

Right heart catheterization mainly helps measure shunt flow and pulmonary artery pressure. Jiang et al[6] report 6 cases of the ratio of pulmonary flow to systemic blood flow (Qp/Qs) value ranging from 1.12 to 1.4, indicating that this deformity does not produce much shunt flow. In these cases, their pulmonary artery pressure was 14 to 30 mmHg, and in 5 cases of EF>62% was 20 mmHg or less. In one patient with heart failure, who had an EF of 45% and moderate mitral valve regurgitation, pulmonary artery pressure reached 30 mmHg. These results suggest that patients with normal left ventricular function do not have high pulmonary artery pressure.

In this patient, the chest X-ray did not show important characteristic signs, with mainly increased pulmonary blood, enlarged left ventricle, and convex pulmonary artery segment indicating left to right shunt change. In patients with heart failure and mitral valve regurgitation, X-rays show cardiac enlargement, pulmonary congestion, a convex pulmonary artery segment, etc., which are more commonly seen in infants and young children.

Infants have atypical symptoms, and the only common characteristic is a Grade 2 systolic murmur at the left edge of the sternum, which results from the large amount of collateral blood flow between the left and right coronary arteries. The ECG might show signs of myocardial ischemia, with ST-segment elevation and/or pathological Q waves. However, the ECG may be normal in children and adults. Echocardiography, ascending aorta angiography or selective coronary angiography can be used to confirm the diagnosis. In terms of treatment, symptomatic infants can be given digitalis, diuretics, or medications that reduce afterload, and surgery can be performed at 4 years old[3]. Surgical intervention might consist of simple left coronary artery root ligation or that procedure may be combined with a variety of surgical coronary revascularizations. The latter includes the left coronary artery-left subclavian artery anastomosis, saphenous vein bypass graft, coronary artery re-implantation, and intrapulmonary tunnel procedure, and the latter two methods are more appropriate[4]. If severe myocardial injury persists and collateral blood supply is insufficient, any treatment effects will be poor. In the present case, the patient gradually developed rich and large collateral circulation which improved early abnormal ECG with disappearance of ST segment elevation in standard leads II, III, augmented limb lead aVF, precordial leads V_5, and V_6, whereas left ventricular hypertrophy, and ECG T-wave inversion in leads V_{1-3} remained. Simple left coronary artery root ligation was performed 6 months later, and the patient was in good condition at 1-year follow-up with no myocardial ischemia.

In summary, in younger patients who have heart murmur and ischemic ECG changes, the possibility of an ALCAPA should be considered. Echocardiography can provide important clues for the diagnosis, and aortic root angiography or coronary angiography can be used to confirm it[7-8].

References

[1] Askenazi J, Nadas AS. Anomalous left coronary artery originating from the pulmonary artery. Report on 15 cases[J]. Circulation, 1975,

51(6): 976-987.

[2] Mollet NR, Cademartiri F, van Mieghem CA, et al. High-resolution spiral computed tomography coronary angiography in patients referred for diagnostic conventional coronary angiography[J]. Circulation, 2005, 112(15): 2318-2323.

[3] Zheng JY, Li H, Chen Y, et al. Anomalous left coronary artery arising from the pulmonary artery discovered beyond infancy[J]. J Clin Ultrasound, 2016, 44(4): 261-264.

[4] Tkebuchava T, Carrel T, Von Segesser L, et al. Repair of anomalous origin of the left coronary artery from the pulmonary artery without early and late mortality in 9 patients[J]. J Cardiovasc Surg, 1991, 33(4): 479-485.

[5] Duan X, Yu T, Wang F, et al. Anomalous origin of the left coronary artery from the pulmonary artery in infants: imaging findings and clinical implications of cardiac computed tomography[J]. J Comput Assist Tomogr, 2015, 39(2): 189-195.

[6] 蒋雄京, 刘国仗, 凌坚, 等. 冠状动脉异常起源于肺动脉六例[J]. 中华心血管病杂志, 2000, 28(1): 74.
JIANG Xiongjing, LIU Guozhang, LING Jian, et al. Anomalous origin of coronary artery from the pulmonary artery: Six-case report[J]. Chinese Journal of Cardiology, 2000, 28(1): 74.

[7] De Kleijn MC, Kuijpers SH, Meijboom FJ. Anomalous left coronary artery arising from the pulmonary artery in an adult women[J]. Neth Heart J, 2016, 24(11): 691-692.

[8] de Leval MR. Comprehensive surgical management of congenital heart disease[J]. J Royal SocMed, 2004, 97(8): 407-408.

(Edited by CHEN Liwen)

本文引用： 周利平, 李小刚, 祁敏, 王爱民. 左侧冠状动脉起源于肺动脉患者心脏骤停成功救治1例[J]. 中南大学学报(医学版), 2017, 42(12): 1458-1464. DOI:10.11817/j.issn.1672-7347.2017.12.016

Cite this article as: ZHOULiping, LI Xiaogang, QI Min, WANG Aimin. Successful treatment in a patient with sudden cardiac arrest due to anomalous left coronary artery arising from pulmonary artery[J]. Journal of Central South University. Medical Science, 2017, 42(12): 1458-1464. DOI:10.11817/j.issn.1672-7347.2017.12.016