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Successful Repair of Infant with Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery Presenting with Heart Failure, Dilated Cardiomyopathy and Mitral Regurgitation

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Abstract
Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital heart anomaly that account for up to 0.5% of all congenital heart disease. Usually, infants' patients may present with failure to thrive, feeding difficulties, sweating, irritability or sudden cardiac death. However, they may be asymptomatic. This syndrome has a high mortality rate if left untreated during infancy period. Echocardiography and electrocardiography are helpful in detecting and establishing the diagnosis of Anomalous origin of the left coronary artery from the pulmonary artery for infants. Surgical repair to restore normal anatomical position of coronary system is the definitive intervention. Early detection and surgical intervention has significant positive impact on survival rate. This paper presents a case of a four months old girl, who presented with heart failure, dilated cardiomyopathy and mitral regurgitation and thereafter was diagnosed with Anomalous origin of the left coronary artery from the pulmonary artery syndrome.
Keywords: Anomalous origin of the left coronary artery from the pulmonary artery- Transthoracic echocardiography -Left anterior descending artery- Surgical repair.

1. Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) or Bland-White-Garland Syndrome is a rare anomaly that account for up to 0.5% of all congenital heart disease, which still could be an underestimation because some patients die before they are diagnosed (1). This condition arises from abnormal morphogenesis of coronary artery during fetal life (2). Clinical presentation occurs when pulmonary vascular resistance and pulmonary arterial pressure drop after birth. This will lead to abnormal flow to left coronary artery or even reversal blood flow leading to myocardial ischemia (3, 4). There are two types of ALCAPA, infants and adults' types. In infants, the symptoms usually start appearing within eight weeks after birth. In contrast, patients with ALCAPA adult type may be asymptomatic or may be presented with mitral insufficiency, ischemic cardiomyopathy, or malignant dysrhythmias, which lead to sudden death (5). Infants with ALCAPA present with feeding difficulties, irritability, vital signs abnormalities, sudden cardiac death, cardiomyopathy and mitral insufficiency that are caused by myocardial ischemia (1, 6). Presence of collateral blood supply to the left coronary artery may contribute to milder or even no symptoms in childhood (6). If untreated, the outcome in ALCAPA syndrome will have extremely high mortality rate (around 90 %) during first year of life (7). Early recognition and surgical intervention to restore the normal blood flow is essential for treatment of ALCAPA patient (8). In this manuscript, we report a case of a four months old girl, who presented with heart failure, dilated cardiomyopathy and mitral regurgitation and diagnosed to have ALCAPA syndrome.

2. Case Presentation

A 4-months old girl was referred because of fatigue, decreased activity, shortness of breath, sweating during feeding with interrupted feeding and choking with no cyanosis. Her symptoms started at 2 months of age. Her medication were furosemide and captopril. On clinical examination her height was 62 cm and she weighed 4.37 kg with pulse of 127 per minute, blood pressure 89/55 mmHg and respiratory rate 42 breath/min. Respiratory examination was within normal limits and breath sounds were equal bilaterally. Cardiovascular auscultation revealed S4 with systolic murmur of mitral regurgitation. Electrocardiography revealed voltage criteria of left ventricular hypertrophy and T wave inversion in inferior lead that indicate myocardial ischemia (figure 1A). Transthoracic echocardiography documented anomalous left coronary artery from pulmonary artery with retrograde flow, dilated right coronary artery seen arising normally from the aorta, echo dense mitral chordae and papillary muscle with shortening of the chordae noted due to ischemic changes, thickened mitral valve leaflets with mildly increased peak velocity of 1.3m/sec (7mmHg) and mean velocity of 0.9m/sec (4mmHg), moderate mitral regurgitation with no systolic flow reversal seen in pulmonary veins, severely dilated left atrium and left ventricle, severely depressed left ventricular systolic function (figure 2B). Based on these finding, this patient was diagnosed with ALCAPA syndrome. Consequently, the patient schedule for surgical repair. The patient underwent left coronary artery transfer to the aorta. Post-operative transthoracic echocardiography

showed good repair with improving function, therefore, the patient was discharged. On follow up, the patient's cardiac function showed significant improvement of ventricular contraction by transthoracic echocardiography with ejection fraction 67.8 % (figure 3C).

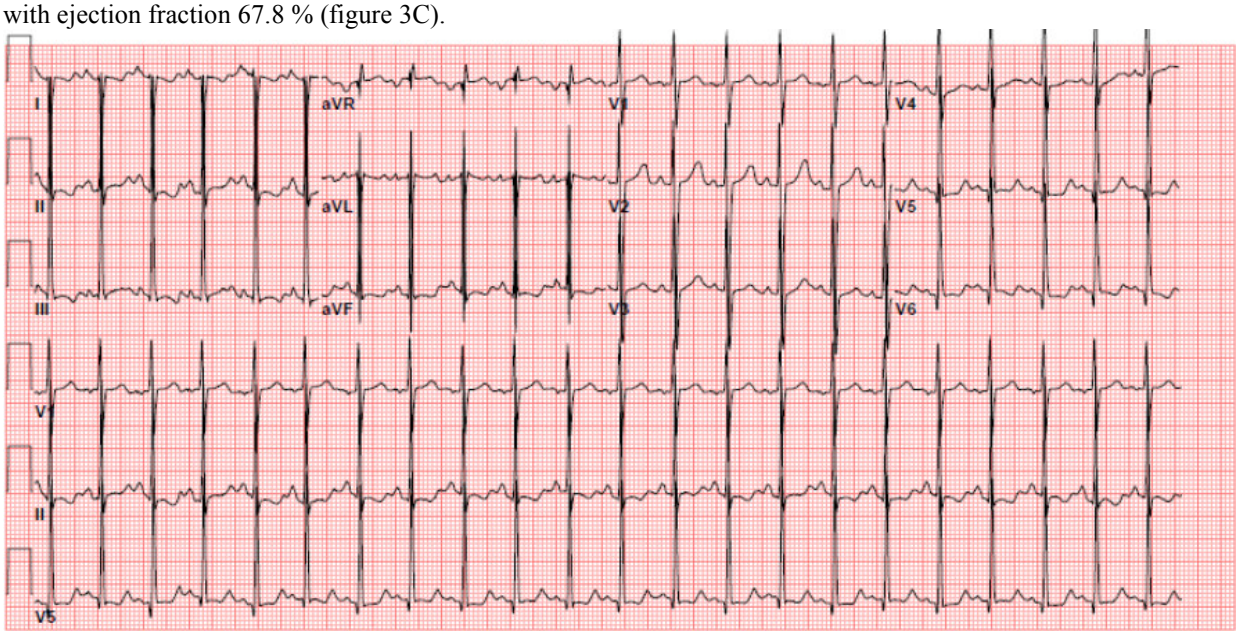

Figure 1A, Electrocardiography showed voltage criteria left ventricular hypertrophy and T wave inversion in the inferior lead

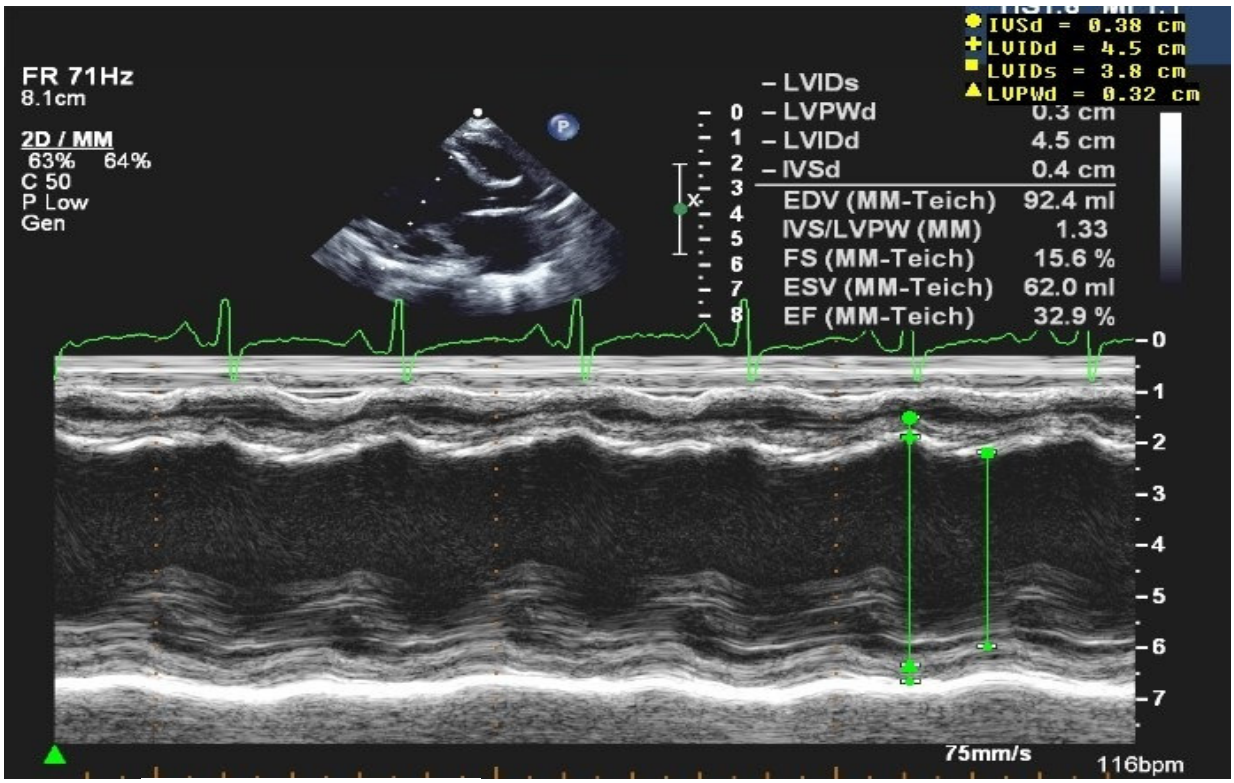

Figure 2B, Preoperative echocardiography showed dilated cardiomyopathy with depressed left ventricular systolic function

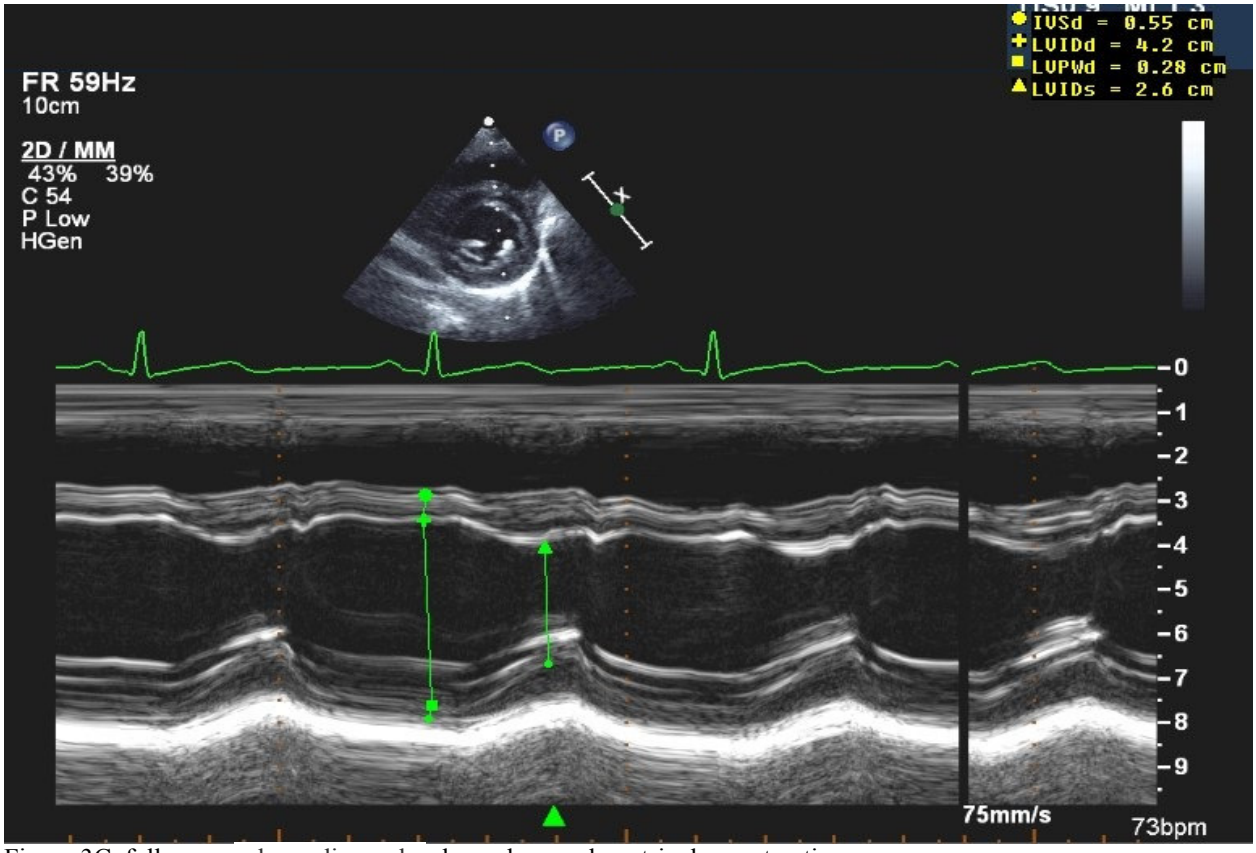

Figure 3C, follow up echocardiography showed normal ventricular contraction

3. Discussion

ALCAPA is a rare congenital cardiovascular anomaly that should be treated surgically as soon as diagnosis is made. This condition is asymptomatic in utero due to equal oxygen saturation and blood pressure in pulmonary artery and aorta (9). However, this will differ after birth. They can present with failure to thrive, feeding difficulties, sweating, irritability or sudden cardiac death due to cardiac muscles ischemia (1, 6). They could survive until adulthood with smaller stature, angina like symptoms, reduced effort tolerance, syncope or even sudden cardiac death (9). Patients may present with myocardial infarction, left ventricular dysfunction, mitral regurgitation, or silent myocardial ischemia, leading to sudden cardiac death (10). Therefore, ALCAPA syndrome must be suspected among infants with cardiomyopathy because it is has high mortality during first year of life and is curable if treated. Even though ECG is usually showing abnormal findings, normal reading of ECG does not rule out the suspicion of ALCAPA (11). The diagnosis of ALCAPA on echocardiography is often simple. However, it can be missed (11). When ventricular dysfunction, mitral regurgitation and dilated cardiomyopathy seen in infants, ALCAPA must be suspected (11). In contrast, coronary *computed tomography* angiography considers the first modality for diagnosis adult type of ALCAPA syndrome (12). Imaging modalities are challenging to sonographers because of the tidal wave of improvement in techniques and medical knowledge. From 2004 to 2007, about 50,660 echocardiograms were reviewed in Boston Children's Hospital and showed the prevalence of misdiagnosing ALCAPA was 16% of echocardiograph (13). Misdiagnosing of ALCAPA may lead to life-threatening clinical consequences (14). In this patient, electrocardiography showed voltage criteria of left ventricular hypertrophy as well as T wave inversion in the inferior lead that indicated an ischemia. In addition, dilated cardiomyopathy, moderate mitral regurgitation and retrograde flow from left anterior descending artery (LAD) to main pulmonary artery were seen in echocardiography. Based on these findings, the patient underwent surgical repair of ALCAPA. Normal anatomical position of coronary artery system was re-created successfully by a modified technique to combine aortic and pulmonary flaps (15) Takeuchi repair technique carries an excellent prognosis but complications must be expected regarding this technique like pulmonary artery narrowing, baffle obstruction and right ventricle outflow obstruction (12). However, using this surgical technique is not usually applicable for adult due to age related changes in dimension and strength of vessels (16). Post-operative long-term survival in infants undergoing surgical ALCAPA correction is 94.8 % (17). The patient presented here had uneventful hospital course with no complication. The patient tolerated the procedure well and showed both ventricular and mitral valve function in good condition. Series of echo were done post operatively and showed good repair. The prognosis is good if the

condition discovered early especially during infancy period (17).

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