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# Anomalous Left Coronary Artery From The Pulmonary Artery (ALCAPA) as a Cause of Heart Failure

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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**Conflict of interest:** None declared

**Patient:** Female, 6-month-old
**Final Diagnosis:** Anomalous left coronary artery from the pulmonary artery (ALCAPA)
**Symptoms:** Dyspnea • failure to thrive • feeding problems
**Medication:** —
**Clinical Procedure:** Ultrasound
**Specialty:** Cardiology

**Objective:** Rare disease
**Background:** Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare anomaly. When present it can result in failure to thrive and congestive heart failure.
**Case Report:** We present the case of a 6-month old female whose presentation was one of failure to thrive. Point of care ultrasound and electrocardiogram (ECG) were used to diagnose heart failure with consideration of ALCAPA. These tools helped to expedite transfer to a tertiary care center for definitive therapy.
**Conclusions:** Although a rare anomaly, ALCAPA induced heart failure can be quickly identified on bedside ultrasound. Together with ECG findings, the Emergency Physician can expedite the diagnosis and proper disposition.

**MeSH Keywords:** Bland White Garland Syndrome • Heart Failure • Pediatrics • Ultrasonography

**Full-text PDF:** https://www.amjcaserep.com/abstract/index/idArt/917655





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## Background

We present the case of a 6-month old female presenting in respiratory distress who was ultimately diagnosed with congestive heart failure secondary to anomalous left coronary artery from the pulmonary artery (ALCAPA). There are 2 types of ALCAPA, infant and adult. The infant form presents at around 8 weeks of age with symptoms of ischemic cardiomyopathy. During this time, the infant develops a drop in pulmonary vascular resistance, and results in infarction in the left ventricle (LV). If left untreated, 90% of patients die within several months after birth. The adult form presents later in life after the formation of collateral vessels in the coronary circulation. These collaterals do not provide enough oxygen to the left ventricle. This chronic hypoxic state can cause malignant arrhythmias, and 80% to 90% of these patients present with sudden death. Our patient in the Emergency Department had a delayed presentation of ALCAPA. The diagnosis of heart failure was expedited using bedside point of care ultrasonography and the differential of ALCAPA was considered after interpretation of the electrocardiogram (ECG) (Table 1). The final diagnosis was confirmed by formal echocardiogram and direct visualization during catheter placement for extracorporeal membrane oxygenation (ECMO).

## Case Report

Our patient presented with breathing difficulties in the setting of upper respiratory symptoms for about one month. She was initially evaluated by her primary care provider and was thought to have an upper respiratory illness. Respiratory syncytial virus (RSV) and flu testing were negative. When her tachypnea persisted, a clinical diagnosis of pneumonia was made. She showed no improvement after an empiric course of antibiotics. At the time of the fourth visit, a chest x-ray was ordered. Cardiomegaly demonstrated on this radiograph prompted referral to the Emergency Department (ED).

At the time of the ED visit, the patient's mother reported cough and congestion, decreased formula feedings, and decreased urine output. Her mother stated, "She had always done this weird breathing thing while feeding," described as a grunting noise. The patient had lost 1-pound over the past month. Vital signs on presentation were heart rate bpm (beats per minute) 150s to 170s, respiratory rate (breaths per minute) 50s to 60s, blood pressure 105 to 84/61 to 31 mmHg, temperature 36.7°C, and pulse oximetry 96% on room air. She was diaphoretic, with subcostal retractions, nasal flaring and grunting. An enlarged liver 2 cm below costal margin was palpated. The child was placed on positive pressure ventilation immediately (CPAP) with improvement in respiratory rate and work of breathing.

**Table 1.** Summary of Echocardiography and ECG findings confirming ALCAPA.

| |
|---|
| **Echocardiography: short axis aortic root view** |
| Large origin of RCA evident from usual coronary cusp |
| LCA origin from aortic root not evident |
| Linear tracking of LAD/CX coursing to a main LCA arising from posterior aspect of MPA |
| Extensive network of collateral flow from RCA connections distally |
| **Electrocardiogram findings** |
| Biatrial enlargement |
| Biventricular hypertrophy |
| Deep Q waves in AVL, V4–V6 |

RCA – right coronary artery; LCA – left coronary artery; LAD – left anterior descending artery; LCX – left circumflex artery; MPA – mid-pulmonary artery.

An electrocardiogram showed sinus tachycardia at 130 bpm, increased precordial voltage and concern for biventricular hypertrophy (Figure 1). Diagnostic evaluation also included a repeat chest radiograph (Figure 2) which demonstrated cardiomegaly. A bedside ultrasound (Figure 3) demonstrated grossly decreased ejection fraction (EF) with a dilated left ventricle (LV) and a floppy mitral valve on color flow doppler but no pericardial effusion. Relevant screening laboratories included a beta-natriuretic peptide > 4500 pg/mL and a troponin I of 0.19 ng/mL.

The patient was transferred to a children's hospital. Soon after arrival, she had a bradycardic arrest thought secondary to an aspiration event. She was intubated and return of spontaneous circulation was achieved (ROSC) after 7 minutes. She was then placed on ECMO. She underwent definitive surgical repair of her ALCAPA from the pulmonary artery by direct implantation into the aorta. The patient was then decannulated 72 hours after ECMO support. Her left ventricle (LV) systolic function was depressed and required Milrinone infusions post-operatively. The patient was discharged home on digoxin, as well as enalapril and furosemide. She had poor weight gain with breast feeding and required feedings via nasogastric tube to promote weight gain. Two months post-operatively, the patient was doing very well.

## Discussion

ALCAPA is a rare disease, occurring in 1 in 300 000 live births [1] that if untreated causes heart failure, myocardial ischemia, and death. It was first described clinically in 1866 but was not



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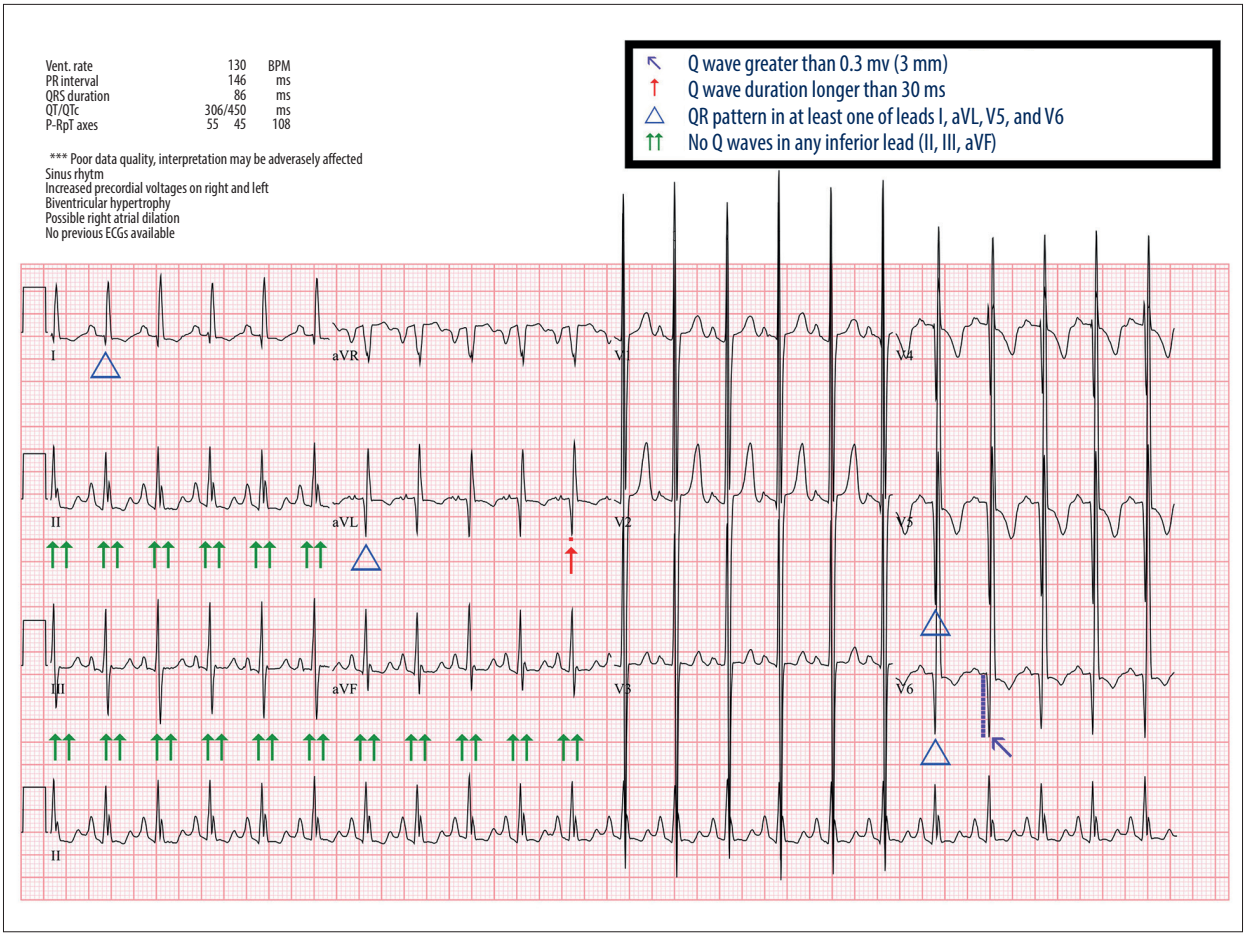


**Figure 1.** Electrocardiogram (ECG) demonstrates sinus tachycardia at 130 beats per minute, appropriate for age. Normal axis and intervals. Increased Q wave duration and Q wave greater than 0.3 mv. QR pattern in at least one of leads aVL, V5, and V6 and absence of Q waves in inferior leads II, III, and aVF consistent with ALCAPA [6,7].

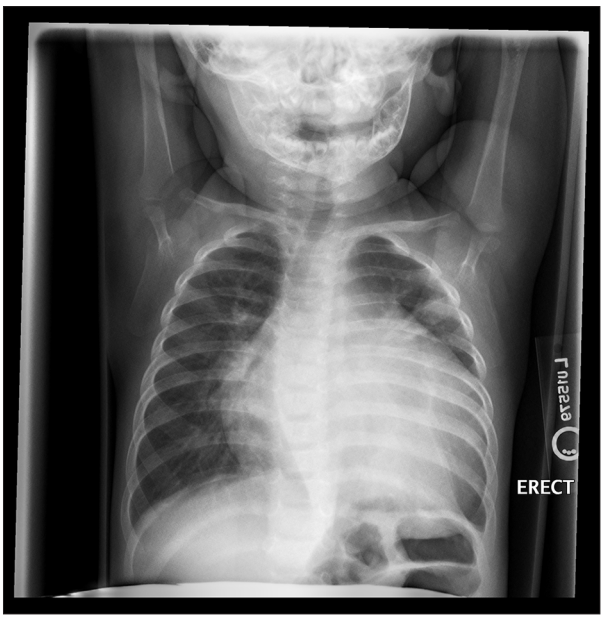


**Figure 2.** Anteroposterior (AP) view of chest radiograph. Cardiac silhouette is significantly enlarged, differential included pericardial effusion or cardiomegaly. Lungs are clear, without pleural effusion, focal pneumonia, or pneumothorax. Soft tissues and bony structures are otherwise unremarkable.

reported on autopsy until 1933 by Bland-White-Garland [2,3]. In normal fetal development, both coronary arteries originate from the aorta. Anomalies can occur during fetal development with abnormal septation of the 2 great vessels or by involution of the aortic buds. When the left coronary artery (LCA) originates from the pulmonary artery (PA), the left ventricle (LV) receives deoxygenated blood. The surrounding arteries start to create collateral blood flow to the LV. At the same time, the blood flow is reversed in the LCA because the PA is a low-pressure vessel compared to the arterial collaterals. This is described as coronary steal syndrome.



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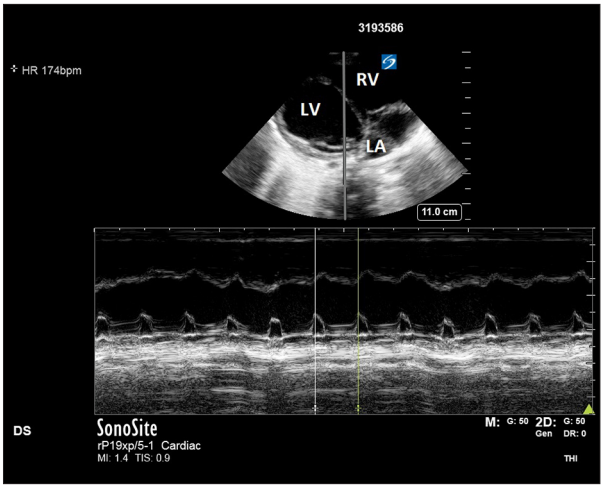


**Figure 3.** Point-of-care cardiac ultrasonography with parasternal long axis view. Left ventricle (LV) is dilated. M-mode shows a widened E point septal separation (EPSS), distance between the white and yellow X marks is 16.0 mm, consistent with a decreased ejection fraction [8].

Patients experience ischemic symptoms during times of increased stress to the heart. In infants this occurs during feedings. Patients may initially present with subtle symptoms of extreme fussiness with feeds, due to the ischemia, and ultimately progress toward respiratory distress as heart failure ensues. Treatment of ALCAPA requires surgical reimplantation of the LCA to the normal anatomical position on the proximal aorta [4,5].

The 2 aspects of diagnosing heart failure in infancy are: 1) chronology of the symptoms and 2) age of the patient. Our patient was older than the typically aged patient that would present with ALCAPA in infancy. Other diagnostic considerations for our patient, and any infant with left ventricular failure, are: ventricular septal defect, dilated cardiomyopathy, valvular disease, infective endocarditis, and myocarditis [9].

Ischemia in the pediatric patient is very rare. Once you have evidence of ischemia, in general, the diagnosis of ALCAPA moves much higher on the differential diagnosis.

## Conclusions

ALCAPA requires surgical repair and has a high mortality if not identified. Point-of-care ultrasound can quickly confirm heart failure in these children; and, if they have characteristic ECG findings, they should be referred to a facility with pediatric cardiothoracic capabilities.

### Conflicts of interest

None.

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