An 8-Month-Old With a Chronic Cough

Teresa M. Whited, DNP, APRN, CPNP-PC, &
Deena D. Garner, DNP, APRN, CPNP-PC

KEY WORDS
Cardiology, chronic cough, failure to thrive, ALCAPA

INTRODUCTION

Cough is a common and chief complaint in primary care. Chronic cough, which is defined as cough lasting for a duration of 4 or more weeks by the American College of Chest Physicians Guidelines (Chang et al., 2017), requires medical attention; it is important to determine the etiology of chronic cough in infants and children. Although chronic cough can be commonly attributed to airway disorders and infectious diseases, it is important to consider less-common diagnoses and use appropriate pediatric guidelines when diagnosing and treating the underlying cause (Bergamini, Kantar, Cutrera, & Italian Pediatric Cough Interest Group, 2017; Chang et al., 2017). A consideration of rare diagnoses such as vascular rings, cardiomyopathies, congenital heart defects (e.g., double aortic arch), cystic fibrosis, and bronchiectasis is needed when the patient does not respond to the prescribed treatment regimen. The following case report illustrates an unusual cause of chronic cough in an 8-month-old infant requiring urgent cardiac surgery.

Teresa M. Whited, Director of MNSc Program, College of Nursing, University of Arkansas for Medical Sciences, Little Rock, AR.

Deena D. Garner, Specialty Coordinator of PNP Program, College of Nursing, University of Arkansas for Medical Sciences, Little Rock, AR.

Conflicts of interest: Teresa M. Whited and Deena D. Garner both receive speaker fees for the NAPNAP Primary Care Review course; they have no other conflicts of interest to report.

Correspondence: Deena D. Garner, DNP, APRN, CPNP-PC, College of Nursing, University of Arkansas for Medical Sciences, 5301 W. Markham Street, #529, Little Rock, AR 72205-7101; e-mail: ddgarner@uams.edu.
J Pediatr Health Care. (2020) *34*, 63-66

0891-5245/$36.00

Published online July 12, 2019.

https://doi.org/10.1016/j.pedhc.2019.06.007

CASE PRESENTATION

An 8-month-old infant presented to Pediatric Cardiology with a chief complaint of chronic cough and possible cardiomegaly on a chest x-ray (CXR). The child had been seen in the pediatric clinic the week before and was found to have a worsening cough with wheezing and rhonchi, for which a CXR was ordered. The radiology report indicated possible cardiomegaly as opposed to poor inspiratory effort. Based on the concern for cardiomegaly, the primary care provider referred the child to cardiology. The child's mother reported that her child had suffered from a chronic cough, which, while fluctuating in severity, had persisted since the child was 2 months of age. She stated that the child had been treated for upper respiratory infections and allergies, with albuterol and fluticasone inhalers for 3 months with no improvement.

In addition, the mother stated that the child had been a poor feeder since she was approximately 2 months old, with frequent bouts of crying during feedings, although calming after eating. She was prescribed ranitidine at 3 months of age for gastroesoph`ageal reflux. Her mother remarked that the infant seemed to be less active compared with her siblings. During routine well-child visits, she was found to have met all developmental milestones, with the exception of being small for her age (less than the fifth percentile for weight). A murmur was not documented by her primary care provider.

The past medical history was significant for term delivery without pregnancy or birth complications. The patient had no previous hospitalizations or surgeries. Family medical history was significant for a maternal great-aunt and maternal great-grandfather with hypertension and a maternal grandfather with early coronary artery disease, but it was otherwise unremarkable. Social history was significant for maternal tobacco smoke exposure; however, the mother reported smoking only outside.

In the cardiology office, the vital signs included a heart rate of 125 beats/min, a respiratory rate of 50 breaths/min, blood pressure of 92/56 mm Hg, and oxygen saturation of 100% on room air. In general, the patient was small for her age (5th percentile for weight, 25th percentile for length)

and appeared thin but was in no acute distress. Respiratory examination revealed intermittent and diffuse crackles bilaterally coupled with a wet cough. Cardiac examination revealed a Grade II/VI pan-systolic murmur, best heard over the left lower sternal border toward the apex, with a point of maximal impulse at the fifth intercostal space lateral to the midclavicular line. Her abdominal examination revealed no masses or tenderness, with hepatomegaly appreciated 3 cm below the right costal margin at the midclavicular line. The remainder of the physical examination was unremarkable.

The patient underwent an electrocardiogram, which showed a deep Q wave in V_1, and a repeat CXR revealed significant cardiomegaly and pulmonary edema. The echocardiogram showed severe dilation of the left ventricle with depressed function and an ejection fraction of 10%, with possible anomalous coronary artery and mitral regurgitation. The child also underwent cardiac catheterization, which confirmed the diagnosis of anomalous left coronary artery arising from the pulmonary artery (ALCAPA), severely dilated left ventricle with depressed function, and moderate mitral regurgitation with paradoxical septal motion.

Following definitive diagnosis by catheterization, the patient was transferred to a pediatric cardiac transplant center over concern that she may require a left ventricular assist device or transplant, if left ventricular function did not improve with surgical repair. The patient underwent a reimplantation of anomalous left coronary artery to the aorta. The postoperative course was relatively uneventful other than a small pericardial effusion, which was managed with anti-inflammatory and steroid medications. After repair, the patient had mild mitral regurgitation with a persistently dilated left ventricle and poor function.

The patient underwent an electrophysiology study approximately 6 months after her surgery for identification of possible sustained ventricular arrhythmias. The study showed easily inducible sustained ventricular tachycardia within multiple foci in the right and left ventricles, and an epicardial internal cardiac defibrillator was placed in her abdomen. Within 1 year of placement, the child had experienced multiple short runs of ventricular tachycardia, which required one shock.

The patient has shown improvement of her left ventricle dilation. She remains stable on carvedilol with improvement in ventricular function, only mild depression of function, and trace to mild mitral regurgitation. Carvedilol, a β-blocker, promotes improved function, contractility, and arrhythmia control in patients with cardiomyopathies. She has shown improvement in her growth parameters and has had resolution of her cough and feeding difficulties since her surgical repair.

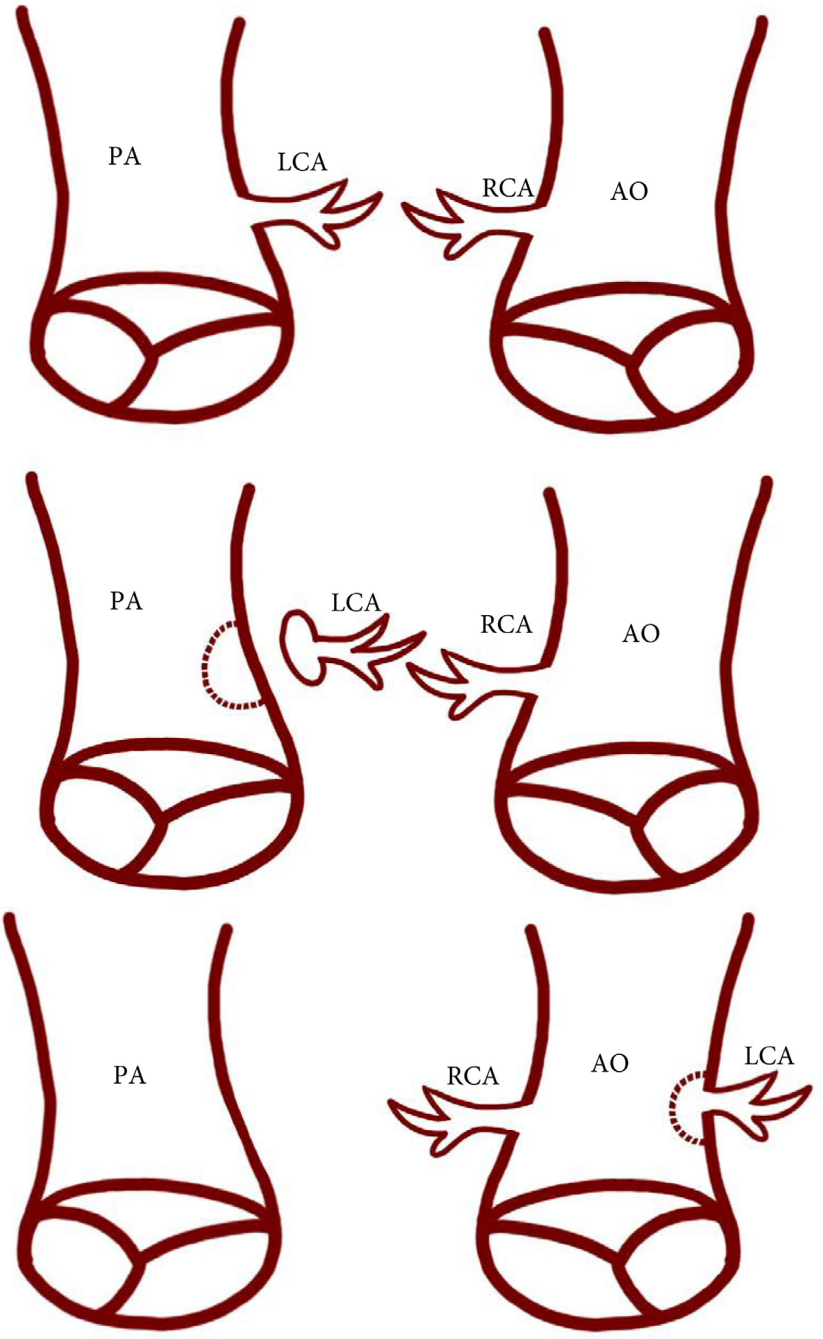
CASE DISCUSSION

The 8-month-old infant in this study had signs and symptoms of ALCAPA, which were indicative of heart failure. The pediatric nurse practitioner should evaluate any cough lasting longer than 4 weeks (including time with or without medication trials for potential underlying cause) with a CXR (Chang et al., 2017). Most infants with ALCAPA are diagnosed around 2 months of age because of a drop in pulmonary vascular resistance, which normally occurs within the first 4–8 weeks of life. The drop in pulmonary vascular resistance results in coronary insufficiency, which leads to symptoms associated with ALCAPA (Weigand, Marshall, Bacha, Chen, & Richmond, 2015). The symptoms of ALCAPA typically include sudden attacks of irritability, especially during feedings, that correlate to episodes of chest pain and symptoms of heart failure such as poor feeding, chronic cough secondary to pulmonary edema, poor weight gain, sweating, pallor, and shortness of breath (Patel, Frommelt, Frommelt, Kutty, & Cramer, 2017). Failure to diagnose ALCAPA within the first 3 months of age places the child at a risk of sudden cardiac death, persistent cardiomyopathy, arrhythmias, cardiac transplant secondary to long-term cardiac malfunction, and mitral valve dysfunction. Untreated infants surviving their first year develop collateral vessels and/or pulmonary hypertension, which allows them to tolerate the poor coronary perfusion associated with ALCAPA, with symptoms arising during times of increased cardiac output such as exercise, feedings, and illness (Tsuda, 2017).

ALCAPA, also known as Bland–White–Garland syndrome, is a rare congenital cardiac anomaly affecting 1 in 300,000 live births (Talwar, Jha, Choudhary, Gupta, & Airan, 2013). In the normal heart, the left coronary artery originates from the aorta and supplies oxygen-rich blood to the muscles of the left side of the heart and the mitral valve. In children with ALCAPA, the left coronary artery originates from the pulmonary artery, which carries oxygen-poor blood from the heart to the lungs (DłużNiewska et al., 2017). During times of physical exertion, such as feedings or exercise, these infants and children are predisposed to myocardial ischemia resulting in myocardial infarction of the left ventricle and the papillary muscles controlling the mitral valve (Patel et al., 2017). Long-term ischemia results in a left ventricular dilated cardiomyopathy and a poorly functioning mitral valve. ALCAPA typically occurs as a single defect but can occur with other congenital cardiac lesions. It is the most common cause of myocardial ischemia and left ventricular infarction in childhood. Myocardial infarction, mitral valve dysfunction, and congestive heart failure are likely to occur within a child's first 3 months of life as a result of poorly oxygenated blood and abnormally low pressures in the coronary arteries. Unless diagnosed and treated within the first year of life, patients face a mortality rate of 90% (Tsuda, 2017).

The evaluation for ALCAPA should include an electrocardiogram, which will show a deep Q wave in V_1, and an echocardiogram, which can identify the coronary anomaly, dilated and poorly functioning left ventricle, and mitral regurgitation. Definitive diagnosis of ALCAPA has historically been made using cardiac catheterization because it allows the identification of the anomaly as well as the assessment of intracardiac and pulmonary pressures (DłużNiewska et al., 2017). Recently, noninvasive imaging techniques such as computed tomography and magnetic resonance imaging have been reported as effective in establishing accurate diagnosis and providing coronary artery morphology information before surgical intervention

FIGURE. ALCAPA with repair. AO, aortic artery; LCA, left coronary artery; PA, pulmonary artery; RCA, right coronary artery.



(This figure appears in color online at www.jpedhc.org.)

(DłużNiewska et al., 2017). Survival is dependent on early recognition of the subtle symptoms of ALCAPA and surgical intervention within the first 2–3 months of life.

During the repair of ALCAPA (Figure), the patient is placed on cardiopulmonary bypass. The anomalous coronary artery is removed using a button technique from the excised pulmonary artery. The proximal coronary artery, with a surrounding button of tissue, is then implanted through direct anastomosis into the aortic root. The pulmonary artery is repaired with autologous pericardium at the site of the coronary removal (Yuan et al., 2018). Most patients tolerate the procedure well when it is performed within the first 2–3 months of life. However, patients with long-standing cardiomyopathy or ischemia (greater than 1 year), and those exhibiting worsening left ventricular function, are at a risk of increased morbidity and mortality and may require prolonged use of inotropic medications and ventilatory support with a prolonged postoperative recovery (Weigand et al., 2015).

In the long term, patients with ALCAPA must be monitored frequently for the recovery of ventricular function, mitral valve function, stenosis of coronary artery graft, and the presence of arrhythmias. Patients with ischemic heart disease resulting from ALCAPA undergo an electrophysiology study to evaluate them for the risk of arrhythmias such as ventricular tachycardia or ventricular fibrillation. According to the American Heart Association/American Colleges of Cardiology/Heart Rhythm Society (2018) guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death, patients with ischemic heart disease, including sustained ventricular tachycardia or unstable ventricular tachycardia/ventricular fibrillation should receive internal cardiac defibrillator placement to prevent sudden cardiac events (Al-Khatib et al., 2018). Such evaluative measures are especially important in young patients such as the 8-month-old infant of this study who presented with long-standing ischemia of the left ventricle. With early detection, timely repair, and long-term management, children with ALCAPA show improvement in ventricular function and have good survival rates.

The diagnosis of ALCAPA can be challenging and may result in delayed referral for treatment in the primary care setting. Gastroesophageal reflux disease, infantile colic, formula intolerance, and respiratory infection are most frequently associated with symptoms such as cough, irritability, and poor feeding. These common conditions are not associated with atypical symptoms of chronic cough, poor weight gain, diaphoresis, and/or pallor. When these prolonged and atypical symptoms and past health history are considered simultaneously, the pediatric nurse practitioner must consider the possibility of ALCAPA or other congenital heart disease.

SUPPLEMENTARY MATERIALS

Supplementary material associated with this article can be found in the online version at https://doi.org/10.1016/j.pedhc.2019.06.007.

REFERENCES

Al-Khatib, S. M., Stevenson, W. G., Ackerman, M. J., Bryant, W. J., Callans, D. J., Curtis, A. B., . . . Page, R. L. (2018). 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: Executive summary: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Heart Rhythm, 15*(10), e190–e252.

Bergamini, M., Kantar, A., Cutrera, R., & Italian Pediatric Cough Interest Group. (2017). Analysis of the literature on chronic cough in children. *Open Respiratory Medicine Journal, 11*, 1–9.

Chang, A. B., Oppenheimer, J. J., Weinberger, M. M., Rubin, B. K., Weir, K., Grant, C. C., . . . CHEST Expert Cough Panel. (2017). Use of management pathways or algorithms in children with chronic cough: CHEST guideline and expert panel report. *Chest, 151*(4), 875–883.

DłużNiewska, N., Kostkiewicz, M., Tomkiewicz-Pająk, L., Krupiński, M., Podolec, J., Olszowska, M., & Podolec, P. (2017). Noninvasive techniques for the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery in adult patients. *Acta Cardiologica, 72*(1), 41–46.

Patel, S. G., Frommelt, M. A., Frommelt, P. C., Kutty, S., & Cramer, J. W. (2017). Echocardiographic diagnosis, surgical treatment, and outcomes of anomalous left coronary artery from the pulmonary artery. *Journal of the American Society of Echocardiography, 30*(9), 896–903.

Tsuda, T. (2017). Preclinical coronary artery anomalies and silent myocardial ischemia in children: How can we identify the potentially life-threatening conditions. *Journal of Pediatric Cardiology and Cardiac Surgery, 1*(1), 49–60.

Talwar, S., Jha, A. J., Choudhary, S. K., Gupta, S. K., & Airan, B. (2013). Repair of anomalous left coronary artery from pulmonary artery (ALCAPA) beyond infancy. *Heart Surgery Forum, 16*(4), E210–E215.

Weigand, J., Marshall, C. D., Bacha, E. A., Chen, J. M., & Richmond, M. E. (2015). Repair of anomalous left coronary artery from the pulmonary artery in the modern era: Preoperative predictors of immediate postoperative outcomes and long term cardiac follow-up. *Pediatric Cardiology, 36*(6), 489–497.

Yuan, X., Li, B., Sun, H., Yan, Y., Meng, H., Xu, L., . . . Xu, J. (2018). Surgical outcome in adolescents and adults with anomalous left coronary artery from pulmonary artery. *The Annals of Thoracic Surgery, 106*(6), 1860–1867.