Review Paper:

Anomalous Origin of Left Coronary Artery From Pulmonary Artery: A Case Series and Review of Literature

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Citation Ghaemi HR, Zamani H, Babazadeh K, Navaeifar MR. Anomalous Origin of Left Coronary Artery From Pulmonary Artery: A Case Series and Review of Literature. Journal of Pediatrics Review. 2020; 8(2):127-132. http://dx.doi.org/10.32598/jpr.8.2.127

http://dx.doi.org/10.32598/jpr.8.2.127

Article info:
Received: 16 Jun 2019
First Revision: 27 Aug 2019
Accepted: 07 Sep 2019
Published: 01 Apr 2020

Key Words:
Anomalous left coronary artery from the pulmonary artery (ALCAPA), Children, Cardiac, Bland-White-Garland syndrome, Coronary vessel anomalies, Cardiovascular diseases

ABSTRACT

Introduction: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare cardiovascular disease presented with an incidence of 1 per 300000 live births.

Case Presentation: In this article, we present four cases of ALCAPA in infancy. Two infants presented with respiratory distress and two with a heart murmur. Their coronary arteries were derived from their pulmonary arteries. Two of them died, one after surgical procedure and one before surgical intervention. Of remained cases, one of them had a good surgical outcome, and another was missed in the follow-up.

A brief review was done on case reports of ALCAPA in children. Eighteen articles were found, including 201 pediatric cases.

Conclusions: The vast majority of affected children will die within the first year of life if ALCAPA is not diagnosed and treated appropriately.

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1. Introduction

Congenital coronary artery anomaly is a defect in one or more of the coronary arteries of the heart, which may be related to the origin, structure, and course of these arteries (1). Its incidence varies between 0.2% and 5.6% in the general population (2). Anomalous Left Coronary Artery From The Pulmonary Artery (ALCAPA) is a heart defect presented in 0.25%-0.5% of the children (3) with an incidence of 1 per 300000 live births (4, 5).

The vast majority of affected children, up to 90%, die within the first year of life if ALCAPA is not diagnosed and treated on time (3, 4). This anomaly was first described anatomically by Brooks in 1885 (6). Bland, White, and Garland described the first clinical features with an autopsy finding of ALCAPA in 1933 (7). This anomaly has thus been named the Bland-White-Garland syndrome (8).

2. Case Presentation

In this study, we report 4 pediatric cases of ALCAPA in the north of Iran, Mazandaran Province, from March 2011 to 2016. The informed consent was taken from each cases' parent.

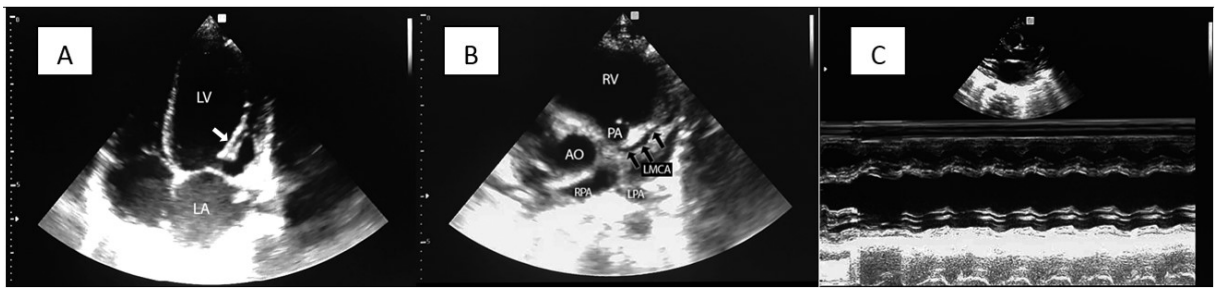
Case 1

A 2-day-old boy was admitted to the neonatal intensive care unit due to respiratory distress and meconium aspiration. Cardiology consultation was performed on the fifth day of admission. Echocardiography showed severe Right Ventricle (RV) and Left Ventricle (LV) dysfunction in addition to pulmonary hypertension. His Tricuspid Regurgitation (TR) gradient was equal to 50 mm Hg, and there was a right to left shunt through a small atrial septal defect. The patient underwent heart failure treatment. During admission, barium swallow was performed due to frequent vomiting episodes, which revealed severe gastro-esophageal reflux.

In the second echocardiography, which was performed 5 days later, his RV function improved and TR was 40 mm Hg, but the LV function was still low. In echocardiography, his left coronary artery was connected to the pulmonary artery, but the diastolic flow was not detectable in the pulmonary artery because of its hypertension (Figure 1). The infant was referred to the heart surgery center with a primary diagnosis of ALCAPA. In the follow-up, the diagnosis was confirmed with CT-angiography, then the infant was operated, but unfortunately, he died immediately after surgery.

Case 2

A 3-month-old girl was admitted to the pediatric intensive care unit due to fever, respiratory distress, pneumonia, and cardiomegaly on the chest radiogram. Cardiac consultation was requested due to cardiomegaly. The first emergency echocardiography revealed severe cardiac dysfunction and dilatation of the left ventricle, and the Ejection Fraction (EF) was equal to 15%. Due to fever, cardiomegaly, severe cardiac dysfunction, and elevated troponin level, the patient underwent cardiac failure treatment with a primary diagnosis of myocarditis, and also intravenous immunoglobulin was prescribed. She was intubated and in the second echocardiography on the next day, the left coronary to pulmonary artery connection was detected. Because of the patient's general condition, she could not be transferred to the cardiac surgery center. After the improvement of pneumonia and extubation, in the third echocardiography, a mild diastolic flow was detected in the pulmonary artery. Six days after the admission, she was extubated, and on the next day, following an episode of vomiting, she underwent cardiopulmonary arrest and expired.



Journal of Pediatrics Review

Figure 1. Case 1: Echocardiography, A; LV enlargement and increased density of papillary muscle (white arrow), B; Short axis view, LMCA (black arrows) from PA, C; Low LV ejection fraction.

LV: left ventricle; RV: right ventricle; LA: left atrium; LMCA: left main coronary artery; RPA: right pulmonary artery; LPA, left pulmonary artery.

Case 3

A 7-month-old male infant was referred to the cardiac clinic due to Failure to Thrive (FTT) and a heart murmur. Echocardiography showed dilated cardiomyopathy, EF=20%, and mitral insufficiency. A careful evaluation revealed the connection of the left coronary artery to the pulmonary artery and distinct retrograde flow in the left coronary artery. The condition was described for the parents, and admission was advised, but they did not refer and follow-up evaluation was impossible maybe due to his referring to another center.

Case 4

A 35-day-old boy was referred to the cardiac clinic due to cardiac murmur. The left ventricle function was decreased in echocardiography, EF=40%, and moderate mitral insufficiency was detected. Evaluation of coronary arteries showed the connection of the left coronary artery to the pulmonary artery, and distinct diastolic flow and reversal flow were not detected in the left coronary artery. The patient was referred to a cardiac surgery center with an initial diagnosis of ALCAPA. In the follow-up, ALCAPA was confirmed, and the patient was operated successfully. The patient is still followed up.

3. Discussion

In fetal life, the pressure of the pulmonary artery is equal to the systemic pressure, allowing for enough myocardial perfusion from the pulmonary artery derived from the anomalous coronary artery (4). However, after birth, the pulmonary artery carries desaturated blood at a pressure that rapidly falls below systemic pressure (9). Hence, the left ventricle is perfused with desaturated hemoglobin at low pressure. This condition predisposes the heart's muscle to ischemia, especially during activities like feeding or crying (4).

After birth, as the resistance of pulmonary arteries decreases, the flow in the left coronary artery and the collateral, tends to pass into the pulmonary artery rather than into the myocardial blood vessels because the pressure in the pulmonary arteries is lower than the coronary arteries. So, a "coronary artery steal" takes place from the coronary arteries into the pulmonary artery (10). This steal phenomenon further leads to myocardial ischemia, and the ischemia worsens during activities such as feeding and crying (4).

Heart failure may occur because of myocardial infarction in the anterolateral region and or mitral valve dysfunction due to ischemia of anterolateral papillary muscle (11). The heart enlarges, and congestive heart failure often worsens by myocardial infarction in the anterolateral region and mitral valve dysfunction secondary to a dilated mitral ring or infarction of the papillary muscles (11).

The ALCAPA syndrome has two types: the adult type and the infantile type. Patients with good collateral vessels have the adult type of ALCAPA, and those without well-established collateral vessels have the infantile type. The manifestations and outcomes of these two types of ALCAPA are different (12). ALCAPA is often isolated but may be associated with other anomalies, including patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot, pulmonary atresia, hemitruncus, and Coarctation of the Aorta (CoA) (3, 4, 11).

The affected infants experience pneumonia and heart failure in the early weeks and months after birth (13). Dyspnea, feeding intolerance, diaphoresis, and FTT are common symptoms that may resemble those of infantile colic, gastroesophageal reflux, and bronchitis (4).

Infants have episodic attacks of restlessness which are equivalent to angina pectoris (11). In infancy and early childhood, ALCAPA is characterized by heart failure symptoms (14). While in late childhood and adolescence, mitral insufficiency (due to papillary muscle infarction), and sudden cardiac death are common manifestations of the disease (14). Dilated Cardiomyopathy (DCM) is an important differential diagnosis for ALCAPA, and this vascular anomaly should be considered in all children with DCM or isolated mitral insufficiency (11).

Distinct cardiomegaly and evidence of pulmonary edema are seen in the chest radiogram (4, 13). Abnormal Q wave and inverted T in the leads I and AVL and V4-V6 are found in EKG, which can help the diagnosis (13). Also, 2D echocardiography and color Doppler are diagnostic and angiography is not necessary (4). Color Doppler evaluation shows the retrograde flow from an aberrant left coronary artery into the pulmonary artery (4). Other findings that help the diagnosis are hyper-density of the mitral papillary muscles, increased septal collateral flow in color Doppler, and significant right coronary artery dilatation (13). Other diagnostic modalities include CT angiography and MRI (4).

ALCAPA is a rare disease that needs a cardiac operation, but in the case of inappropriate treatment, its mortality reaches up to 90% within the first year of life (3). Of four reported cases in this study, 2 cases expired. One of them died before the surgery and another one im-

mediately after the operation, one case is alive now, and we miss follow up of the latter one.

Review protocol

A quick review was made on online databases for articles in the English language that reported ALCAPA cases in pediatric age. We searched the keywords of "children", "pediatric", "ALCAPA", "Anomalous left coronary", and "Bland0-White-Garland syndrome" in the Google Scholar and PubMed without time limitation. Finally, we found 18 published case reports with 201 cases (Table 1).

Zheng et al. reported 19 cases during 16 years (12 boys and 7 girls). The age range of them was 2.5 months to 13 years, an average of 12 months. Operation mortality was 5 (26%) patients (13).

In Uysal et al. study for 20 years, 7 ALCAPA cases were reported, and 5 of them were girls. In 5 patients, the clinical manifestation was heart failure and all of them aged less than 6 months, and two of them were referred due to heart murmur (15).

In the case series of Brotherton et al. (11) 5 patients were reported for 5 years, 3 girls and 2 boys, and all of them were under 5 years old. In our case series for 5 years, 4 ALCAPA cases, 3 boys and 1 girl, were reported and all of them aged less than 1 year old and 2 of them were neonates.

Table 1. Pediatric cases of ALCAPA

Study, Year	Ref.	Location	Cases	Male No. (%)	Prognosis
Ma, 2017	(17)	China	3	2 (67)	All had post-operative good outcome.
Zhang, 2017	(18)	China	50	29 (58)	Overall, the midterm follow-up showed satisfactory recovery of cardiac function after the successful restoration of a dual-coronary arterial system.
Walker, 2016	(19)	The United States	1	0 (0)	Post-operative good outcome
Moeinipour, 2016	(12)	Iran	5	4 (80)	All had post-operative good outcomes.
Rodriguez, 2015	(20)	The United States	12	2 (33)	The overall survival rate was 83%; in the older, asymptomatic children, it was 100%
Muzaffar, 2014	(21)	India	53	29 (55)	There were 5 postoperative hospital deaths with an overall mortality rate of 9.6%
Molaei, 2014	(22)	Iran	2	0 (0)	No follow up
Aliku, 2014	(23)	Uganda	1	1 (100)	Died before operation
Uysal, 2013	(15)	Turkey	7	2 (28)	Three cases have died. Two of these patients died during the post-operative period, and the remaining patient died suddenly during the preoperative period at home.
Szmigielska, 2013	(24)	Poland	1	1 (100)	Good postoperative outcome
Smith, 2013	(25)	The United States	1	1 (100)	No follow up
Secinaro, 2011	(5)	The United kingdom	6	3 (50)	No follow up
Zheng, 2010	(13)	China	19	12 (63)	In addition to the 5 perioperative deaths, 13 patients survived post-operatively without overt symptoms during the follow-up period.
Ojala, 2009	(3)	Finland	29	--	Early postoperative mortality (<30 days) was 17%. No late mortality (>30 days) was detected.
Brotherton, 2005	(11)	Ireland	5	2 (40)	Three had good postoperative outcomes, one diagnosed post mortem, and one died after the operation.
Bland, 1993	(8)	The United States	1	1 (100)	diagnosed post mortem
Salzer, 1993	(26)	Austria	3	2 (67)	No follow up
Brooks, 1885	(6)	Ireland	2	--	No follow up

Some cases of adulthood ALCAPA were reported by Fierens et al. (16) (a 73-year-old woman) and Selzman et al. (a 23-year-old woman) (14).

Symptoms of the disease in infancy are various and can resemble common complications of infancy, including colic, gastroesophageal reflux, lactose intolerance, and pneumonia. The ALCAPA diagnosis can be mistaken by dilated cardiomyopathy and endocardial fibroelastosis (13). In Ojala et al. study for 27 years, 29 cases were reported, of which 4 patients were admitted with the initial diagnosis of pulmonary infection (3).

In our reported patients, an infant was diagnosed due to pneumonia and cardiac insufficiency. In another neonate, the ALCAPA was diagnosed due to cardiac insufficiency and another one due to a heart murmur and FTT. One infant was diagnosed due to the detection of a heart murmur.

For diagnosis of ALCAPA, 2D, and color echography are diagnostic, and in color Doppler, retrograde flow in the pulmonary artery is observed (4). In this case series, in the first case, the aberrant coronary artery was detected in 2D echocardiography, but due to some degrees of pulmonary hypertension, reversed flow was not detected in the pulmonary artery.

In the second case, the aberrant coronary artery was detected too, and the reversed flow was not observed in the pulmonary artery. Some days later, following recovery from pneumonia and extubation, a mild reversed flow was detected in the pulmonary artery.

A clear inverted flow was reported in the third case, who was a 7-month-old infant who is justified according to the age of the infant and decreased pulmonary hypertension.

In the first and second cases, ALCAPA was diagnosed in the second echocardiography, presumably due to heart failure and precision in the connection of coronary arteries. All of our cases had some evidence of heart failure and lacked any other congenital heart disease.

4. Conclusions

Although rare, in infants and children presented with dilated cardiomyopathy, decreased heart function, or myocardial infarction, ALCAPA should be considered as an important differential diagnosis, and the connection of coronary arteries to aorta should be carefully checked in echocardiography.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles were considered in this article. The parents of the patient provided informed written consent for the release of results and data.

Funding

The study did not receive any financial support.

Authors contributions

Conceptualization and design: Hamid Reza Ghaemi and Mohammad Reza Navaeifar; Review of data: Kazem Babazadeh; Drafting of the manuscript: Hassan Zamani; Critical revision of the manuscript for important intellectual content: Hamid Reza Ghaemi.

Conflict of interest

The authors declared no conflict of interest.

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