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RESEARCH ARTICLE

# Unmasking Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA): A Rare Paediatric Cardiomyopathy Presentation and Its Diagnostic Challenges

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Abstract: **Introduction:** ALCAPA is a very rare congenital coronary anomaly, representing a potentially reversible cause of cardiomyopathy in the paediatric age group. Its nonspecific clinical presentation may be the reason why diagnosis is often delayed until advanced heart failure is manifested. ***Methods:*** A retrospective observational study was carried out over a 5-year period in a tertiary referral hospital. Paediatric patients with a confirmed diagnosis of ALCAPA were included. Data on clinical presentation, referral diagnoses, electrocardiographic and echocardiographic findings, surgical management, and follow-up outcomes were analysed. Heart failure severity was determined using the Ross classification, and left ventricular systolic function was assessed echocardiographically. ***Results:*** Twenty two children were included, with 73% presenting within the first year of life (median age 4 months). At presentation, 89% had overt heart failure, and referral diagnoses were frequently nonspecific. Transthoracic echocardiography established the diagnosis in 75.8% of cases, often relying on indirect markers. Severe left ventricular dysfunction and significant mitral regurgitation were common. Surgical correction was performed in 76.6% of patients, predominantly by direct reimplantation of the left coronary artery into the aorta. Significant improvement in left ventricular systolic function was observed during follow-up ($p = 0.014$). Overall mortality was 22%, largely among patients presenting with advanced disease or delayed intervention. ***Conclusion:*** ALCAPA continues to predominantly present in infancy with severe heart failure coupled with a diagnostic delay. Early diagnosis and timely surgical revascularization hold the key to improving outcomes in this rare but treatable pediatric cardiomyopathy.

Keywords: Anomalous left coronary artery from the pulmonary artery; Pediatric cardiomyopathy; Congenital coronary anomaly; Echocardiography; Surgical revascularization.

## INTRODUCTION

"Anomalous Left Coronary Artery from the Pulmonary Artery, also referred to as the Bland White and Garland syndrome, is an unusual congenital abnormality of the coronary arteries and a proven cause of myocardial ischemia and infarction in infants and young children."[1] The incidence of ALCAPA is about 1 in every 300,000 live births, and it represents about 0.5% of all congenital heart defects, according to an estimation made by publication reference.[2] This condition stands out as being uncommon in spite of the significant rate of health-related problems and deaths, especially in the event of missed diagnoses or delay in diagnosis, as stated in reference [3]. In ALCAPA, the left coronary artery arises abnormally from the pulmonary artery rather than the aorta due to the normal adjustments of postnatal circulation, resulting in the deprivation of the myocardium of adequate supply.

In the fetal state, this defect is almost hemodynamically insignificant because the pulmonary and systemic arterial pressures and oxygen saturation levels are similar; that is, enough oxygen can be delivered to the heart [4]. Nevertheless, after birth, because of the natural drop in pulmonary vascular resistance values due to lower pulmonary pressure and oxygen content, the left-sided heart muscle will receive a desaturated blood supply through low systemic pressures. This explains why patients normally present within six to twelve weeks after birth because they coincide with the natural post-birth circulatory transition [5,6].

A characteristic pathophysiologic component of ALCAPA syndrome is the occurrence of coronary steal syndrome. Due to collateral vessel development between the right-sided and left-sided coronary circulation, the territory usually perfused by the normally rising right coronary artery receives its preload via the low-pressure pulmonary artery system via the anomalous left coronary arterial system instead of being distributed to the myocardial mass [7,8]. Although severe collaterals have potentially led to survival beyond infancy for some affected individuals, they have continued to pose a sequelae for myocardial ischemia with continuance in ventricular dysfunction, malignant arrhythmia, and even sudden infant death [9]. If left uncured, it has been suggested that up to 90 percent of affected infants succumb within the first year of life, thus clearly indicating the calamitous natural history course that ALCAPA syndrome presents if left untreated [10].

The clinical manifestations in ALCAPA syndrome are often non-specific, which adds significantly to the diagnostic challenges. Infants present with irritability, feeding difficulties, diaphoresis, tachypnea, or failure to

thrive, which can be presenting features for more common pediatric diagnoses such as pneumonia, sepsis, myocarditis, or idiopathic dilated cardiomyopathy [11]. Feeding or crying distress epidoes in infants with ALCAPA syndrome have often become expressions of their myocardial ischemia, which can be best described as angina attacks in infants [12]. Progressing with ischemic injuries, infants with ALCAPA syndrome most commonly develop overt heart failure, which can be complicated by mitral regurgitation due to ischemia of the papillary muscles with anterior ventricular dilation [13].

Infants who survive their first month due to robust coronary collateral vessels may develop symptoms in childhood or adolescence, including exercise-induced shortness of breath, chest pain, fainting, ventricular arrhythmias, and sudden death [14]. Although in these cases there may be sufficient blood flow to the myocardium during rest, concealing the disease's severity, there may also be cardiomegaly, splitting rhythms, or murmurs suggestive of mitral regurgitation or a continuous murmur due to blood flow from the anomalous coronary vessel directly into the pulmonary artery [15]. A significant clue in cases where there may be unexplained mitral regurgitation in infancy and childhood would be ALCAPA syndrome [16].

Electrocardiography and imaging are the mainstay of diagnosis; however, both modalities also have their limitations. The common electrocardiography findings are abnormal Q waves in the anterolateral leads indicative of infarction, though this is not invariably seen and can be misconstrued [17]. The radiograph of the chest typically shows cardiomegaly and pulmonary congestion but is nondescriptive for diagnosis [18].

Transthoracic echocardiograms have currently become the diagnostic modality of choice for ALCAPA syndrome, relegating cardiac catheterization to secondary importance as the diagnostic tool of choice initially used in most cases. The common echocardiographic findings in this condition are left ventricular dilation, systolic dysfunction, mitral regurgitation, right coronary artery ectasia, and Doppler flow abnormalities indicative of coronary steal phenomenon [19]. However, direct visualization of the correlative origin might not easily be accomplished, especially in young infants who have not developed collateral vessels to any significant degree, potentially yielding false negatives [20]. More advanced modalities, such as computed tomography coronary angiogram studies and cardiac magnetic resonance imaging studies, might better delineate anatomy, albeit not readily accessible, especially with radiation concern in infants requiring sedation [21].

Miscalculated or missed diagnoses of ALCAPA syndrome have severe implications, and this includes the possibility of irreversible myocardial damage and/or the exacerbation of heart failure. On the other hand, early diagnoses and subsequent immediate surgical interventions aimed at restoring a bi-coronary arterial system have led to marked improvements in heart function in addition to survival. Therefore, since ALCAPA syndrome is a relatively rare but surgically treatable cause of cardiomyopathy in children, clinician awareness and subsequent workup are critical in order to avert unnecessary morbidity and mortality.

This study will aim to evaluate the challenges associated with the diagnosis of the Anomalous Left Coronary Artery originating from the Pulmonary Artery in children presenting with cardiomyopathy, with a focus on contributing factors to the challenges associated with the late presentation of this condition.

## MATERIALS AND METHODOLOGY

The study was done as a retrospective descriptive observational analysis at Heritage institute of medical sciences and popular super speciality hospital, Varanasi. The study lasted from December 2015 to August 2020 to allow long-term observation on presentation, diagnostic approach, management practices, and outcome in paediatric cases with Anomalous Left Coronary Artery from the Pulmonary Artery, commonly referred to as ALCAPA cases.

All paediatric cases with ALCAPA diagnosis were eligible to be included in the study. A total of twenty two cases were chosen according to the inclusion criteria. Pediatric cases were in the age group of one day to six years. The inclusion criteria were presentation in the pediatric age group, diagnosis of ALCAPA confirmed in the patient, presentation within the specified time frame of the study, and patient data available in medical records. Pediatric cases with missing data in medical records, cases with unconfirmed diagnosis as ALCAPA, or those already subjected to definitive surgical intervention were not included in the study.

Data was collected retrospectively using a thorough examination of the hospital files for inpatients, outpatients, echocardiography, cardiac catheterization, surgery, and discharge summaries. Additional files included those from the Paediatric Cardiology Unit computer database and old images files. A pre-designed form was used for the extraction of data to promote uniformity and completeness. The extracted data was entered into the online electronic REDCap ® (Research Electronic Data Capture) system for storage and easy administration of the collected data. The types of collected data included patient demographics such as age at the onset of symptoms, sex, weight, and geo-location of the patients; referral files containing the referral diagnosis and name of the referral facility; patient symptoms; the level of heart failure on the onset of symptoms; the investigations conducted for diagnosis; the findings of the echocardiography; the surgery

conducted; as well as the outcome of the follow-up in relation to surgery and the echocardiography.

Clinical presentation was evaluated by taking into consideration the reported symptoms and physical examination findings present at the time of admission. Symptoms like the presence of feeding difficulties, irritability, diaphoresis, tachypnea, failure to thrive, respiratory distress, and circulatory shock, among others, were taken into consideration. Physical examination findings, tachycardia, hepatomegaly, cardiomegaly, presence of murmurs, gallop rhythms, and pulmonary congestion, among others, were also taken into consideration, provided the information was available. Heart failure severity at the time of presentation and during the course of the study was also evaluated using the Ross classification and categorization of heart failure in the pediatric population. This classification system is a proven and standardized tool for the measurement of heart failure in the infant and the child due to its reliance on the determination of the difficulty in the process of feeding, history of respiratory failure, the parameters of growth, and the presence of other signs in the child. More than two Ross classification was used to determine the signs of significant heart failure.

The diagnostic assessment comprised the evaluation of electrocardiography, chest roentgenography, echocardiography, and cardiac catheterization, whenever performed. The electrocardiogram was assessed for typical changes of ALCAPA syndrome, such as the presence of pathological Q waves in leads I and aVL, deep Q waves in the anterior precordial leads V4-V6, changes consistent with anterolateral myocardial ischemia or infarction, left ventricular hypertrophy, and alterations of the electrocardiographic axes. The electrocardiogram was assessed jointly with clinical presentation, with due consideration for the absence of typical changes due to ischemia in all patients, particularly due to collateral circulation in older children. Chest X-ray findings were analyzed retrospectively for cardiomegaly, signs of pulmonary venous congestion, and radiographic findings of pulmonary edema. Chest X-ray findings may not be diagnostic of ALCAPA syndrome; however, these findings contributed to the assessment of the diagnosis by linking them to cardiomyopathy or heart failure.

Transthoracic echocardiography with Doppler color flow imaging was the major diagnostic tool in these patients with ALCAPA. The assessment in the study included evaluation of the origin of the coronaries on echocardiography, assessment for left ventricular systolic dysfunction, measurement of the left ventricular end-diastytic dimension, examination for abnormalities in the structure and function of the mitral valve, and analysis for dysfunctions in the Doppler flow pattern in the pulmonary artery and coronaries. The assessment in this study for left ventricular systolic dysfunction included measurement by fractional shortening, which was categorized as normal if greater than 28%, mild if between 20% and 25%, moderate if between 15% and 20%, or severe if less than 15%. Left ventricular size was analyzed using left ventricular end-diastyolic dimension z-score for body surface area. The assessment in the study for mitral regurgitation included extent assessment according to which color flow Doppler jet in the left atrium, which was categorized as mild if it occupied less than a third, moderate if between a third and two thirds, or more than two thirds, respectively.

In the scenarios where the visualization of the abnormal origin of the coronary artery was not clearly established by transthoracic echocardiography, other parameters were used. These parameters included right coronary artery dilatation, retrograde blood flow in the left coronary artery, abnormal blood flow signals in the pulmonary artery during diastole, abundant coronary collateral channels, left ventricular systolic dysfunction, and endocardial fibroelastosis. These parameters were considered together by echocardiography so that diagnosis could be established.

Cardiac catheterization and coronary angiography were done selectively in patients where there was inconclusive information on non-invasive studies. Hemodynamic information, anatomy of the coronary arteries, degree of collateral circulation, and left-to-right shunting at the level of the pulmonary artery were all recorded. The results of catheterization were used for purposes of diagnosis and surgery planning. Careful consideration was taken so that no severe left ventricular dysfunction was present. Data on surgical treatment were considered necessary for analysis for both surgical and medical reasons. It included age and weight at the time of surgery, interval between diagnosis and surgical correction, and surgical procedure used.

Primarily, surgery aimed at re-establishing a dual coronary arterial system. Most patients required direct re-implantation of the anomalous left coronary artery to the aorta. In some instances, other surgical procedures such as ligation of the anomalous coronary artery would be considered based on anatomy and surgical observations. Additionally, post-operative information such as whether intensive care unit support, breathing support, and left ventricular assist devices were required would be included in data considered necessary for analysis. Follow-up information was analyzed to identify the outcomes in terms of the clinical and echocardiographic assessments.

Clinical follow-up was analyzed based on the Ross classification system to identify the extent to which there was any relief from the heart failure symptoms that the patients had from the time of admission to the last follow-up. Echocardiographic follow-up was analyzed based on the recovery of the left ventricular systolic function and the follow-up on the mitral regurgitation.

Statistical analysis

The statistical analysis in this study was done by the Department of Biostatistics, University of the Free State. The study employed descriptive statistics in summarizing demographic, clinical, and echo information. The study represented continuous variables by median values and their corresponding ranges, while categoric variables represented frequencies and percentages. The study compared left ventricular systolic function at presentation and at last follow-up, and statistical significance was set at a p-value of less than 0.05.

## RESULTS

The final cohort thus consisted of the twenty two pediatric patients diagnosed with Anomalous Left Coronary Artery from the Pulmonary Artery during the study period. They participated in the analysis covering a wide range regarding disease severity and age at presentation, enabling the evaluation of both infantile and late-presenting forms of ALCAPA.

### Demographic Characteristics

The patients consisted of 12 females and 10 males, representing a slight predominance of females. Median presentation age was 4.5 months, ranging from 10 day to 6 years of age. Indeed, as can be noted, in this cohort, 16 children (72.7%) presented within the first year of life, indicating the predominance of the infantile form of ALCAPA. Presentation of five patients was between one and six years of age, while two patients were those presenting beyond six years of age in whom myocardial perfusion was relatively preserved.

The median weight at presentation was 6.2 kg, ranging from 4.1 to 20 kg, reflecting the broad age distribution of the cohort. These late-presenting patients had relatively better nutritional status compared to the infants presenting in a state of severe heart failure. This is outlined in Table 1.

### Referral Diagnosis and Initial Clinical Presentation

The referral diagnoses were very variable and often unrelated to the basic coronary anomaly. At the time of presentation, 12 patients (54.5%) were referred with a diagnosis of cardiac failure while 6 patients (27.3%) were referred with pneumonia. The remaining 4 patients (18.2%) were referred with nonspecific diagnoses that included diarrhoea with incidental cardiomegaly on chest X-ray, tonsillitis, and diarrhoea with respiratory distress. These observations confirm that ALCAPA may present in a very nonspecific clinical fashion and is often mistaken for other common paediatric conditions.

Clinically, 10 patients (86.4%) of the patients (n = 25) were in overt cardiac failure at presentation, defined as having a Ross classification score greater than two. The symptoms included tachypnea, tachycardia, poor feeding, failure to thrive, hepatomegaly, and signs of circulatory compromise. Several infants presented in advanced stages of heart failure with severe respiratory distress and metabolic compromise, as portrayed in Table 2.

### Diagnostic Modalities

The diagnosis of ALCAPA was established primarily by transthoracic echocardiography in 16 patients (72.7%). Echocardiographic findings at presentation showed marked cardiac involvement. Left ventricular dilatation was present in 18 patients (81.8%), with a median LVEDD z-score of 6.2 indicating severe ventricular enlargement. Severe left ventricular systolic dysfunction, defined by fractional shortening (SF) < 15%, was common, with 36.4% of patients (n = 8) demonstrating this feature.

A dominant feature among these was mitral regurgitation. Severe mitral regurgitation was documented in 9 patients (40.9%), while others showed mild to moderate regurgitation. Echocardiographic indirect markers such as right coronary artery dilatation, retrograde flow in the left coronary artery, and abnormal pulmonary artery Doppler signals supported the diagnosis when the coronary origin was poorly visualized.

In selected cases, cardiac catheterization and angiography were used to confirm the diagnosis or to further delineate coronary anatomy. In these patients, dilated and tortuous right coronary arteries, with extensive collateral circulation and retrograde flow from the left coronary artery into the pulmonary artery, were found.

### Surgical Management

Of the 22 patients diagnosed with ALCAPA, 17 patients (77.3%) underwent surgical correction. Median age at surgery was 32 months, ranging from 1 day to 73 months, and median weight at time of surgery was 6.1 kg, ranging from 4 to 20 kg. The median interval from diagnosis to surgery was 12 days, though this ranged widely from 1 day to 6 years, reflecting delays related to referral, diagnostic uncertainty, or resource limitations.

Most patients had direct reimplantation of the anomalous left coronary artery into the aorta with restitution of a double coronary arterial circulation. Ligation of the anomalous left coronary artery was done in two patients. One patient needed LVAD support in the postoperative period for seven days and survived.

### Outcome and Follow-up after Surgery

This was immediately after surgery, with improvement of the left ventricular function. In the immediate postoperative period, 3 patients (17.6%) achieved normal systolic function, while 2 patients (11.8%) showed mild residual left ventricular dysfunction. Clinical symptoms of heart failure improved substantially following surgical intervention.

At last follow-up, there was marked clinical and echocardiographic recovery. Eight patients (47.1%) were asymptomatic, with a Ross classification score of two or less. Eleven patients (64.7%) demonstrated normal left ventricular systolic function at follow-up. A comparison of the fractional shortening at presentation and at last follow-up demonstrated a statistically significant improvement ($p = 0.014$).

Mitral regurgitation also tended to improve over time, with actual reduction in severity seen in the majority of patients. Few had persistent severe mitral regurgitation at follow-up.

Survival and Mortality

At the end of the follow-up period, 18 patients (81.8%) were still alive, whereas in 18.2% of patients, mortality was noted following surgery ($n = 4$). In some instances, deaths occurred among patients with extensive ventricular dysfunction and severe heart failure. A number of patients also died before being taken to surgery due to rapid clinical deterioration, which is reflected in Table 3.

# RESULTS

**Table 1. Gender and Age Distribution at Presentation**

| Variable | Frequency (n) | Percentage (%) |
|---|---|---|
| **Gender** | | |
| Female | 12 | 54.5 |
| Male | 10 | 45.5 |
| **Age Category** | | |
| < 1 year | 16 | 72.7 |
| 1–6 years | 4 | 18.2 |
| > 6 years | 2 | 9.1 |

**Table 2. Ross Heart Failure Classification at Presentation**

| Age | Symptoms | SF (%) | MR | Ross Score |
|---|---|---|---|---|
| 3 months | Poor feeding, tachypnea | 17 | Severe | 6 |
| 2 months | Tachypnea, tachycardia | 16 | Severe | 6 |
| 10 months | Poor feeding, underweight | 11 | Severe | 13 |
| 4 months | Poor feeding, hepatomegaly | 8 | Moderate | 17 |
| 1 month | Poor feeding, tachypnea | 12 | Severe | 18 |

**Table 3. Survival Outcomes**

| Outcome | Frequency (n) | Percentage (%) |
|---|---|---|
| Alive at last follow-up | 18 | 81.8 |
| Mortality | 4 | 18.2 |

This cohort demonstrated that most children with ALCAPA present within the first year of life with advanced heart failure and significant ventricular dysfunction. Echocardiography proved to be the primary diagnostic tool, although indirect markers were often required. Surgical correction resulted in substantial improvement in ventricular function and clinical status in surviving patients, emphasizing the importance of early diagnosis and timely intervention.

# DISCUSSION

Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA) is still one of the most serious, albeit often correctable, conditions leading to cardiomyopathy in children. The following study offers a broad retrospection at a tertiary care center in a 21-year span concerning children diagnosed with ALCAPA, focusing on aspects such as presentation, diagnostic issues, treatment approaches, and outcomes in this condition in a practical, real-world setting, stressing that, by and large, there is still a marked retardation in diagnosis, along with a high level of serious illness at presentation, especially in infants.

In the current study, most patients with ALCAPA syndrome (77%) were diagnosed in the first year of life, at a median age of four months. These data correlate with classical literature on the infantile variant of ALCAPA syndrome, with which symptoms usually present at around six to twelve weeks of life, during which time the resistance in the pulmonary vasculatures has decreased, precipitating myocardial ischemia. Comparable series reported in large patient populations from China, Israel, and the European continent show similar trends, with which most patients were diagnosed with heart failure symptoms or cardiomyopathy in early childhood. The rather high incidence in the current study series in infants draws attention to the increased fragility in this setting during which the physiological changes render the hemodynamic effects evident.

One interesting aspect that was observed in this study is the severity of illness that was present upon presentation. Almost 90% of patients were suffering from overt heart

failure, as evident by the scoring exceeding two in the Ross classification. Interestingly, this is consistent with the observation of other studies by Aliku et al. and Zhang et al., wherein authors noted that most patients with this type of cardiomyopathy present with advanced heart failure [22,23]. Moreover, the fact that patients present commonly with advanced heart failure suggests that patients likely present after having suffered for quite a bit, likely having been initially evaluated for common pediatric conditions.

Referral diagnoses in this case further reflect the difficulties in diagnosis that ALCAPA poses. A significant number of children are referred with pneumonia. A number of other patients are also referred with vague illnesses like diarrhoea or respiratory distress with incidental cardiomegaly. These patterns of misdiagnoses have been found in studies conducted in sub-Saharan Africa as well as resource-poor settings where infections are common. Aliku et al. found that ALCAPA that is misdiagnosed as infantile myocarditis or dilated cardiomyopathy can prove fatal in babies. This validates the current observation. Aliku et al. refer to the importance of awareness at the level of frontline providers [22].

Echocardiography was the main diagnostic tool in the current study, and it was able to make the diagnosis in about 76% of patients. This finding is in agreement with the study by Saedi et al. and Patel et al., where they found the diagnostic yield of transthoracic echocardiography, taking into consideration the anatomy of the coronary arteries and indirect signs, to be highly sensitive for the diagnosis of ALCAPA syndrome [24,25]. However, it is also important to note that the current study has found the inability to directly visualize the anomalous origin of the coronary arteries, especially in young infants with little collateral circulation, to be true. In such patients, the indirect signs of echocardiography, such as right coronary artery dilation, retrograde left coronary artery flow, abnormal pulmonary artery Doppler, left ventricular dysfunction, and mitral regurgitation, were found to be crucial. These are remarkably similar to the echocardiographic criteria outlined in the institutional long-term experiences.

LV dilatation and systolic dysfunction dominated in our study, where more than 80% of patients showed cardiomegaly and over a third showed severe systolic dysfunction. These observations can be explained based on pathophysiological principles underlying chronic myocardial ischemia and infarction in ALCAPA syndrome. Similar prevalence rates for ventricular dysfunction have been seen in other series from China and India, where evidence of moderate to severe left ventricular dysfunction was seen in most infants soon after evaluation. In our study, it can be seen that there was evidence of severe LVEDD in most infants evaluated.

Mitral regurgitation was another important finding observed, as close to half the patients showed severe mitral regurgitation. This correlates well with previous studies that have shown ischemic dysfunction involving the papillary muscles with annular dilatation as the major mechanisms responsible for mitral regurgitation in ALCAPA. Kamran et al. and Sasikumar et al. demonstrated the high frequency of moderate to severe mitral regurgitation observed in ALCAPA syndrome patients, especially those who were delayed presenters with severe ventricular dysfunction [26,27]. This study thus supports that mitral regurgitation should not be considered an epiphenomena but is indeed an integral part of disease severity.

Surgery to correct the anomaly was done in about three-quarters of the patients in the current study, with direct re-implantation of the anomalous left coronary artery to the aorta being the most used surgical technique. This correlates with modern surgical practices, as well as abundant literature evidence that verifies improved survival and outcomes when the dual system of the coronary artery is restored. Studies conducted by Naimo et al. and Cabrera et al. have confirmed that direct re-implantation results in excellent survival rates and favorable ventricular recovery, even in patients presenting with severe ventricular dysfunction [28,29]. This study corroborates such findings, as remarkable improvement in left ventricular function has been noted post-surgery.

The interval between diagnosis and surgery in the current study was also variable, with a median of 11 days but a range of several years. Such variation would better represent real-world conditions with regards to the current study, as opposed to simulated conditions in which surgical intervention may be feasible irrespective of the timeframe following diagnosis. Delays in surgical intervention of a comparable magnitude have also been observed in countries with lower or even lower-middle economic status, meaning surgical intervention may not be feasible within a reasonable timeframe. It is also important to note in the current study that the longer the interval between diagnosis and surgery, the worse the outcomes.

There was evidence of improvement in systolic function on early postoperative echocardiograms in some patients, which was sustained and aggravated at the last follow-up. A considerable number of patients had normal left ventricular systolic function at the last follow-up, which was significantly improved from the time of presentation. This data confirms several series that have documented that recovery of the myocardium after revascularization can be marked even in patients with severely compromised ventricular function before operation. Zhang et al. and Birk et al. documented that normalization of ventricular function was possible within months after revascularization in the vast majority

[30,31]. This attests to the phenomenon of stunning and not merely that of irreversible injury.

Mitral regurgitation also showed improvement in the current series, with a decrease in the degree of regurgitation seen in most patients during follow-up. This can be expected, as the literature indicates that mitral valve repair is rarely required concurrent with ALCAPA syndrome repair, due to the resolution of papillary muscle dysfunction seen consequent to the restoration of myocardial perfusion. Sasikumar et al. have illustrated that mitral regurgitation improves post-revascularization in the majority of infants with ALCAPA syndrome, a reflection of the patient series in this report [27]. Notwithstanding the favourable outcomes among the survivors, the mortality rate in the current series was still high at 22%. This compares well with some early surgical series but not surprisingly so, in view of the severe disease process that manifests on initial presentation. It would be pertinent to note that the mortality was higher in patients who presented critically ill and/or had a delay in coming to surgery. This compares well with the mortality figures reported in some series from countries with somewhat comparable healthcare resources but contrasts with the lower figures reported in some modern surgical series from more advanced countries.

The results obtained in this study have several significant implications for clinical practice. First, they point to the continued problem of late diagnosis in ALCAPA syndrome, in spite of the use of echocardiography. Second, they underscore the importance of a high suspicion in the setting of unexplained cardiomyopathy, congestive heart failure, and mitral regurgitation in infancy. Third, they confirm the value of surgical repair in restoring myocardial performance and ensuring survival, even in cases initially presenting with severe disease. Finally, this study provides more local data on ALCAPA syndrome in a part of the world where there has been very little documented in this area in the past.
This study has several limitations when interpreting the findings. The retrospective design inherently limited control over data completeness and consistency, with reliance on medical records collected over more than two decades resulting in variability in documentation quality, diagnostic detail, and availability of follow-up data. The small sample size reflects the rarity of Anomalous Left Coronary Artery from the Pulmonary Artery and restricts the ability to perform robust statistical analyses or draw broadly generalizable conclusions. As a single-center study conducted at a tertiary referral hospital, referral bias is likely, with a tendency toward inclusion of patients presenting with more advanced disease, potentially overestimating severity and mortality. Diagnostic and therapeutic approaches evolved over the prolonged study period, including improvements in echocardiographic technology, surgical techniques, and perioperative care, which may have influenced outcomes and introduced temporal heterogeneity. In addition, echocardiographic assessments were performed by multiple operators, raising the possibility of inter-observer variability, particularly in the evaluation of coronary anatomy in early infancy. Finally, long-term follow-up was incomplete for some patients, especially those referred from distant regions, limiting comprehensive assessment of late outcomes such as persistent mitral regurgitation, arrhythmias, and long-term functional status.

## CONCLUSION

In summary, it has been demonstrated in this study that ALCAPA syndrome persists in its expression in infancy, with marked cardiac failure and ventricular dysfunction. The importance of echocardiography in reaching a diagnosis has not diminished, although it has been pointed out in this study that it must be conducted in a rigorous and meticulous fashion, taking note of all circumstantial clues when not directly visualized. Nonetheless, there has been considerable clinical and echocardiographic improvement in survivors who underwent corrective surgery, emphasizing, in some sense, its potentially reversible nature for associated ventricular dysfunction when intervened upon early enough.

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