PATIENTS WITH MECHANICAL HEART VALVE THROMBOSIS

The prevalence, characteristics and outcomes of anomalous left coronary artery from the pulmonary artery at the Chris Hani Baragwanath Academic Hospital over a 28-year period

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ABSTRACT

Background: Anomalous left coronary artery from the pulmonary artery (ALCAPA) accounts for 0.25% - 0.5% of congenital cardiac disease. ALCAPA results in myocardial ischaemia and a dilated left ventricle with impaired systolic function which can be reversed post-surgical correction. We describe the presenting clinical features, diagnostic findings (including classical electrocardiographic findings) and post-operative outcomes, including the improvement in left ventricular function, in patients at a South African tertiary care centre.

Methods: A retrospective analysis of patients with ALCAPA over a 28-year period at the Chris Hani Baragwanath Academic Hospital (CHBAH).

Results: A total of 38 patients were included, with 24 (63.2%) females, and a median age at diagnosis of 4.6 months (IQR: 3.2 - 9.1 months). The clinical presentation was variable and included dyspnoea, poor feeding, and a cough. The majority were diagnosed to have a lower respiratory tract infection (71%). Cardiomegaly on chest X-ray (CXR) was present in 84.2% of patients. Deep Q waves in leads I and aVL was the most prevalent finding on electrocardiography in 96.9% of patients. ST segment depression (8 patients) and T wave inversion (21 patients) was evident in the lateral and inferior diaphragmatic leads. Left ventricular ejection fraction (LVEF) improved significantly from 38.8 ± 6.3% to 57.5 ± 9.1% post-surgical correction (p-value=0.0004) at the first follow up (median of 1.3 months). The early mortality rate was 21.6%.

Conclusion: The clinical presentation is often suggestive of a chest infection and cardiomegaly on CXR is common. Specific electrocardiographic features commonly present in patients with ALCAPA may be a guide to making the diagnosis. Surgical correction is associated with improved left ventricular function.

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INTRODUCTION

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a result of a poorly understood disruption in the embryological development of the coronary arteries. ALCAPA is a rare congenital cardiac defect accounting for 0.25% - 0.5% of congenital cardiac disease and occurring approximately once per 300 000 live births.[1] An early consideration of the diagnosis of ALCAPA requires the recognition of a constellation of clinical signs and electrocardiogram (ECG) features, and it is thereafter confirmed with echocardiography.

The clinical findings of ALCAPA are nonspecific and thus necessitates a high clinical index of suspicion. The commonest presenting clinical symptoms and signs found in the literature include dyspnoea, feeding intolerance, failure to thrive, irritability, and the presence of a cardiac murmur.[1-4] Patients may initially be asymptomatic due to a high pulmonary artery pressure supplying the myocardium, albeit at a lower oxygen saturation.[3] As the pulmonary resistance decreases, the pulmonary artery pressure decreases and results in a "coronary steal" phenomenon with subsequent myocardial ischaemia.[3] The presence of a collateral blood supply between the right and left coronary arteries is protective and may influence the timing of presentation as well as the presenting clinical manifestations.

While cardiomegaly is frequently evident on chest X-ray (CXR) in patients with ALCAPA,[3,5] a study by Levitas, et al. demonstrated a normal cardiothoracic ratio (CTR) to be a common finding.[6] More importantly, electrocardiographic (ECG) findings may aid in the diagnosis of ALCAPA. A sinus tachycardia,

abnormally deep Q waves (especially in leads I and aVL), ST segment changes (ST segment elevation or depression) T wave inversion, and poor R wave progression may be seen.[1,2,7,8]

The diagnosis of ALCAPA may be confirmed non-invasively by echocardiography whereby the left coronary artery (LCA) can be seen arising from the pulmonary artery. The right coronary artery becomes the dominant coronary artery and may appear larger than the LCA. Colour Doppler is used to assess flow reversal in the left coronary artery reflective of blood flow from the right coronary artery via collateral vessels to the LCA and then into the pulmonary artery (which is the equivalent of a right to left shunt).[9] The consequences of myocardial ischaemia is manifested by the presence of left ventricular dilatation and dysfunction as well as mitral regurgitation.[2,9] An increased echogenicity of the anterolateral papillary muscle and the adjacent myocardium is a common finding and in keeping with fibrosis and infarction.[9]

An ascending aorta angiography can confirm the diagnosis of ALCAPA.[4] A prominent RCA and with an apparent absent LCA may be seen initially followed by retrograde filling of the LCA via the collateral blood flow from the right coronary artery.[1,2,4,10] Other non-invasive diagnostic modalities such as computerised tomography (CT) scans and magnetic resonance imaging (with the added benefit of no ionising radiation exposure) can also be used to confirm the presence of ALCAPA.

Surgical management is the definitive treatment for ALCAPA. Direct reimplantation of the anomalous coronary artery onto the aorta (DIACA) is the most frequent method used.[1,7,10-12] A less common surgical intervention is the Takeuchi procedure which includes a pulmonary arteriotomy, the creation of an aortopulmonary window and the use of the pericardium to reconstruct the pulmonary artery.[10] Excellent postoperative outcomes have been found in patients with ALCAPA across various studies.[10,11,13] Early mortality rates post-surgery range between zero and 16%, while survival rates of 86% - 100%, 10 years post-surgical repair have been reported.[13]

There is a paucity of data regarding the clinical presentation, diagnostic features and outcomes of ALCAPA in the South African paediatric population. South Africa is a low- to middle-income developing country with a large diverse population, many of whom rely on peripheral hospitals for medical services. The recognition of clinical and electrocardiographic features suggestive of ALCAPA in the secondary care hospital setting would assist in the rapid referral of patients to specialist services for appropriate treatment.

MATERIALS AND METHODS

Data collection

A retrospective analysis of all paediatric patients diagnosed with ALCAPA at the Chris Hani Baragwanath Academic Hospital (CHBAH) between January 1991 and December 2018 was conducted. Thirty-eight patients were identified.

The demographic data, presenting symptoms and clinical features, the initial diagnosis and the CXR features were noted. The ECG was analysed according to age-specific norms. Abnormal Q waves were defined as being more than 25% of the R wave height in the same lead or more than 3mm in lead one or more than 2mm in lead aVL, as well as being more than 0.03 seconds in duration.[1,14] ST segment depression was defined as being more than 1mm below the baseline for more than 0.08 seconds duration.[14] T wave inversion and the leads in which the various ECG changes were observed was documented.

Echocardiographic features of ALCAPA such as flow reversal in the LCA, hyperdensity of the papillary muscles and reduced left ventricular function was documented. The left ventricular ejection fraction (LVEF) was defined as normal if between 56% and 78% (mean 66%).[14] The right coronary artery diameter was interpreted based on the mean body surface area-based normal values.[14] Cardiac catheterisation and angiographic data was analysed if available.

Other data such as the method and timing of surgical repair, post operative duration of follow up, the time to being asymptomatic, normalisation of the LVEF, resolution of the mitral regurgitation and outcomes such as death (pre-, intra-, or post-operatively), and the loss to follow up were documented. Early mortality was defined as being within 30 days of the operation.

Data analysis

All data was captured on a Microsoft Excel spreadsheet and analysed using STATA (MP 13) software. Continuous variables were presented as medians and interquartile ranges (IQR) and categorical variables described using frequencies and percentages. These were compared using a Fisher's exact bivariate analysis, dividing the patients into an infant (patients less than 12 months of age) and a non-infant group (patients more than 12 months of age). The associations between multiple variables and the LVEF at presentation were assessed using a univariate regression analysis. A paired samples t-test was used to compare pre- and post-surgical correction LVEFs.

A Fisher's exact test was used to compare the group of patients who survived to those who demised (pre- and post-surgery), as well as to compare the outcomes over 2 decades (the 1990s and the 2000s). A chi-square test analysis was not feasible due

to the small sample size. It became apparent that a multiple of the expected frequencies in the respective categories consisted of values less than 5. A p-value less than 0.05 was considered statistically significant.

Ethical consideration

The University of the Witwatersrand (WITS) Faculty Graduate Studies Committee, the WITS Human Research Ethics Committee (clearance certificate number: M190741) and the necessary medical authorities at the CHBAH all granted permission to access the study data and conduct this study.

RESULTS

A total of 38 patients were diagnosed with ALCAPA between January 1991 and December 2018 at the CHBAH. This constituted 0.45% of the 8 387 patients with congenital cardiac defects seen by the paediatric cardiology unit over this period.

Age at presentation and anthropometry

The median age of diagnosis was 4.6 months (IQR: 3.2 - 9.1 months), with 24 (63.2%) female and 14 (36.8%) male patients. World Health Organisation (WHO) weight-for-length anthropometry charts showed that 16 (51.6%) patients with available anthropometry data plotted values within the normal range, 3 (9.7%) patients were wasted, and 12 (38.7%) patients were severely wasted on presentation. Twenty-five (65.8%) patients presented to CHBAH initially, while the remaining 13 (34.2%) patients were referred from other medical facilities.

Clinical details at presentation

The majority of patients, 31 (81.6%), had no pre-existing medical conditions at the time of presentation. Four patients presented with other cardiac diagnoses (2 patients with a diagnosis of myocarditis, 1 with an anterior mitral valve leaflet prolapse, and 1 patient had Trisomy 21 with a patent ductus arteriosus) who were then subsequently diagnosed to have ALCAPA. Ten patients (26.3%) were human immunodeficiency virus (HIV) exposed, and 2 (5.3%) were HIV positive on treatment. One patient was being treated for tuberculosis at the time of diagnosis.

The most frequent presenting symptoms (Table I) were dyspnoea in 26 (68.4%), a cough in 16 (42.1%), and a history of poor feeding in 7 (18.4%) patients. The most common clinical findings (Table I) were cardiomegaly, with a displaced apex beat, and respiratory distress both occurring in 20 (52.6%) patients, followed by hepatomegaly in 17 (44.7%) patients.

Majority of the patients (n=27, 71.1%) were initially diagnosed to have a lower respiratory tract infection which is reflective of the non-specific presentation of ALCAPA. The next most common initial diagnoses were dilated cardiomyopathy and myocarditis, each diagnosed in 8 (21.1%) patients. Other cardiac defects were suspected in 3 (7.9%) patients, including a congenital cardiac defect for 2 patients and a patent ductus arteriosus (PDA) in the third patient. Sepsis and congestive cardiac failure were diagnosed in 2 (5.3%) and gastroenteritis in 1 (2.6%) of the 38 patients.

CXR findings

The majority of the patients (n=32, 84.2%) were reported to have cardiomegaly on the initial CXR, while 6 (15.8%) had a normal CTR. The median CTR was 70% (IQR: 61% - 70%). The

TABLE I: The presenting symptoms and clinical features in the 38 patients with ALCAPA at the CHBAH (1991 - 2018).

Variable	Number of patients (% of the total number of patients, n=38)	Variable	Number of patients (% of the total number of patients, n=38)
Presenting symptom		**Clinical feature**	
Dyspnoea	26 (68.4)	Displaced apex beat	20 (52.6)
Cough	16 (42.1)	Respiratory distress	19 (50.0)
Poor feeding	7 (18.4)	Hepatomegaly	17 (44.7)
Gastroenteritis	6 (15.8)	Irritability	11 (28.9)
Diaphoresis	4 (10.5)	Cardiac murmur	8 (21.1)
Fever	3 (7.9)	Tachycardia	7 (18.4)
Cyanosis	2 (5.3)	Tachypnoea	7 (18.4)
Inconsolability	2 (5.3)	Hypotension	4 (10.5)
Shortness of breath	2 (5.3)	Crepitations	2 (5.3)
Fatigue	1 (2.6)	Gallop rhythm	2 (5.3)

most prevalent additional CXR findings included pulmonary congestion in 10 (26.3%) patients, followed by biventricular enlargement in 5 (13.2%) and changes suggestive of bronchopneumonia in 4 (10.5%) patients.

ECG findings

Of the 34 patients with available ECGs at diagnosis (Table II), 29 (85.3%) had a normal heart rate for age, 4 (11.8%) had a tachycardia, and 1 (2.9%) patient had a bradycardia. All of the ECGs showed a sinus rhythm. The majority of patients, 24 (70.6%), had a normal axis for age, 7 (20.6%) had a left axis with the remaining 3 (8.8%) having a rightward axis for age. Deep Q waves (fulfilling the defined criteria) were present in 32 (94.1%) of the ECGs.

The abnormally deep Q waves occurred most frequently in both standard leads I and aVL in 31 (96.9%) patients. ST segment depression occurred in 8 (23.5%) of the 34 patients, most frequently in lead aVF in 4 (50.0%) patients, followed by lead II in 3 (37.5%) and lead aVL in 2 (25.0%) patients. Inverted T waves occurred in 21 (61.8%) patients, most frequently in the lateral and inferior diaphragmatic leads. Of the 83 episodes of abnormal T wave inversion across the 34 ECGs analysed, 37 (44.6%) occurred in the lateral leads: I, aVL, V5 and V6, and 32 (38.6%) in the inferior diaphragmatic leads: II, III and aVF. Additional ECG findings included left ventricular hypertrophy in 17 (50.0%) and biventricular hypertrophy in 4 (11.8%) patients.

Echocardiography findings

Of the 37 patients who had echocardiography studies (Table III), the LCA was visualised originating from the pulmonary artery in 32 (86.5%) patients. The LVEF documented in 36 patients was decreased in 33 (91.7%) patients, with a median LVEF of 34.0% (IQR: 23.5% - 43.5%). Mitral regurgitation was present in 27 (73.0%) patients, with majority graded as mild. An increased echogenicity of the papillary muscles and adjacent endocardium was noted in 26 (70.3%) patients.

Measurement of the right coronary artery diameter was documented in 19 (51.4%) patients, with 18 (94.7%) showing enlargement. Collateral blood flow between the right and left coronary arteries was visualised in 2 (5.4%) patients. Retrograde blood flow from the LCA to the pulmonary artery was evident in 16 (43.2%) patients. Other findings on echocardiography included a PDA, a patent foramen ovale, an anterior mitral valve leaflet prolapse and a left coronary ostial stenosis.

Cardiac catheterisation and angiographic findings

There were 19 (50.0%) patients who underwent cardiac catheterisation and angiography (Table IV). All showed an enlarged RCA originating from the ascending aorta. Retrograde

TABLE II: The electrocardiographic manifestations of ALCAPA.

Variable	Number of patients (% of the total number of ECGs analysed, n=34)
Presence of deep Q waves	32 (94.1)
Leads I and aVL	31 (96.9)
ST segment depression	8 (23.5)
Lead aVF	4 (50.0)
Lead III	3 (37.5)
Lead aVL	2 (25.0)
Inverted T waves	21 (61.8)
Left ventricular hypertrophy	17 (50.0)
Biventricular hypertrophy	4 (11.8)

TABLE III: The electrocardiographic manifestations of ALCAPA.

Variable	Number of patients (% of the total number of patients with echocardiographic findings, n=37)
LCA visualised originating from the pulmonary artery	32 (86.5)
LVEF	36 (97.3)
Decreased	33 (91.7)
Normal	3 (8.3)
Median LVEF = 34.0%, IQR: 23.5 - 43.5%	
Mitral regurgitation	27 (73.0)
Mild	14 (51.9)
Mild-moderate	1 (3.7)
Moderate	7 (25.9)
Moderate-severe	4 (14.8)
Severe	1 (3.7)
Increased echogenicity of the papillary muscles and adjacent endocardium	26 (70.3)
Right coronary artery diameter	19 (51.4)
Dilated	18 (94.7)
Normal	1 (5.3)
Other echocardiographic findings	
Presence of collateral blood flow between the right and left coronary arteries	2 (5.4)
Presence of retrograde blood flow from the LCA to the pulmonary artery	16 (43.2)

filling of the LCA into the pulmonary artery was documented in 16 patients, with 10 (52.6%) patients showing visible collateral blood supply via the right coronary artery.

Surgical correction

Thirty (78.9%) of the 38 patients with ALCAPA underwent surgical correction, with 27 (90.0%) patients undergoing DIACA and in the remaining 3 (10.0%) patients a tunnel was created from the LCA to the aorta. The median number of days to repair from the diagnosis of ALCAPA was 27 days (IQR: 12 - 51 days), with a median age of repair of 5.7 months (IQR: 3.7 - 11.6 months).

Post-surgical follow up findings

Follow up findings were available in 19 (63.3%) patients post operatively (Table V). The median first follow up after surgical repair was 1.3 months (IQR: 0.9 - 1.9 months). Eighteen patients (94.7%) were asymptomatic, and 1 (5.3%) patient was tachypnoeic. A cardiac murmur was found in 4 (21.1%) patients. Other complications noted included a right brachial plexus injury in 1 patient and a right hemiplegia secondary to a left cerebral infarct in another. Both patients had a long intensive care unit (ICU) stay post-surgery. The median time to being asymptomatic was 1.4 months (IQR: 0.9 - 2.0 months).

Of the 18 (94.7%) patients with documented echocardiography data at the first post-surgical follow up, 11 (61.1%) showed a normal LVEF. Ten of these patients initially had a decreased LVEF prior to surgery. The other 7 (41.2%) patients had decreased LVEF at the first follow up post-surgery. The median LVEF was 61% (IQR: 39% - 74%) at the first follow up for these 18 patients. The overall LVEF of these patients improved significantly from (38.8 ± 6.3%) pre-surgical correction to (57.5 ± 9.1%) post-surgical correction (p-value=0.0004). Complete normalisation of the LVEF (Figure 1) occurred at a median of 6 months (IQR: 2 - 10 months) post-surgical correction of ALCAPA.

Of the 18 patients with documented echocardiography at the first follow up post-surgery, mitral regurgitation was initially present prior to surgery in 14 (77.8%) patients, with 6 (50.0%) having moderate to severe mitral regurgitation pre-surgical correction. Mild mitral regurgitation was present in 12 (66.7%) of these patients' post-surgery. Mitral regurgitation resolved at a median of 6 months (IQR: 3 - 9.5 months) post-surgical correction. The median number of months of follow up post-surgery was 121.3 months (IQR: 61.1 - 173.9 months).

TABLE IV: The cardiac catheterisation and angiographic findings in patients with ALCAPA.

Variable	Number of patients (% of the total number of patients with cardiac catheterisation and angiography, n=19)
LCA visualised originating from the pulmonary artery	18 (94.7)
Retrograde blood flow evident from the LCA to the pulmonary artery	16 (84.2)
Presence of collateral blood flow from the right coronary artery	10 (52.6)
Additional findings	
Visibly dilated right coronary artery	4 (21.1)
Left ventricular dysfunction	1 (5.3)
Left ventricular hypertrophy	1 (5.3)
PDA and severe pulmonary hypertension	1 (5.3)

TABLE V: Post-surgery follow up findings.

Variable	Number of patients (% of the total number of patients with post-surgical correction follow up findings, n=19)
Symptoms at the first follow up	
Asymptomatic	18 (94.7)
Tachypnoeic	1 (5.3)
Median months until asymptomatic was 1.37 (IQR: 0.9 - 2 months)	
Echocardiographic findings at the first follow up	18 (94.7)
LVEF (median LVEF of 58% (IQR: 39 - 74%))	18 (100)
Normal	11 (61.1)
Decreased	7 (38.9)
Normalisation of LVEF occurred at a median of 6 months (IQR: 2 - 10 months)	
Mitral regurgitation	12 (66.7)
Grade: mild	12 (100)
Resolution of mitral regurgitation occurred at a median of 6 months (IQR: 3 - 9.5 months)	
Other	
Left ventricular dilatation	3 (16.7)
Pericardial effusion	1 (5.6)
Mild pulmonary regurgitation	1 (5.6)
Aortic regurgitation	1 (5.6)
Residual PDA	1 (5.6)

Outcomes

Outcomes are shown in Figures 2 and 3. At the time of data analysis, 12 (31.6%) patients continued to be followed up at the paediatric cardiology clinic and 2 (5.3%) patients had been transferred to the adult cardiology unit. Eleven (28.9%) patients were lost to follow up, and 8 (21.1%) had demised within 30 days. Three patients demised post-operatively on the day of surgery. The median number of days post-surgery to patient demise was 0.5 days (IQR: 0 - 10.5 days). Three (7.9%) patients demised prior to surgery and 2 (5.3%) absconded prior to surgery. A Fisher's Exact test demonstrated no significant difference in outcomes between patients in the infant vs. the non-infant group (p-value=0.383).

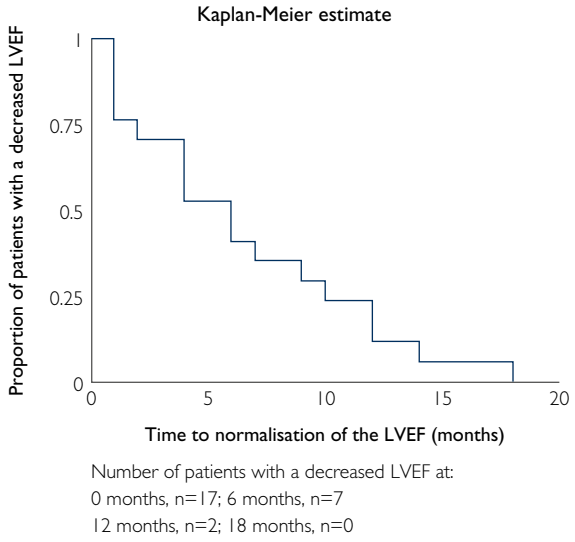

FIGURE 1: Illustrating the time to normalisation of the LVEF following the surgical reimplantation of the LCA.

Additional analysis

Echocardiographic visualisation of the LCA originating from the pulmonary artery and the presence of left ventricular dilatation were statistically significant when comparing the infant and non-infant groups, with p-values of 0.037 and 0.042 respectfully. Analysis of the LVEF and other variables assessed (Table VI) showed that only the presence of anthropometry of the severely wasted category was statistically significant with a p-value of 0.048.

A bivariate analysis of the survival group vs. the non-survival group showed no statistically significant differences with regards to the demographics, diagnostic findings, and the age of repair (Table VII). The median LVEF at diagnosis was 39% (IQR: 30% - 41%) in the survival group versus 31% (IQR: 23% - 40%) in the non-survival group.

A review of the patient cohort over 2 decades – the 1990s and the 2000s - showed no statistically significant differences in the diagnostic modalities between the 2 groups. Mitral regurgitation was diagnosed more frequently in the 2000s group (p-value= 0.001), possibly due to the availability of echocardiographic colour Doppler technology in the more modern echocardiographic machines. The patients in the 2000s group were repaired sooner at a median of 18.5 days (IQR: 9.5 - 46.5 days) vs. the 1990s group which were repaired at a median of 42 days (IQR: 18 - 51 days) post diagnosis. There were no significant differences in outcomes between the 2 groups (p-value=0.126).

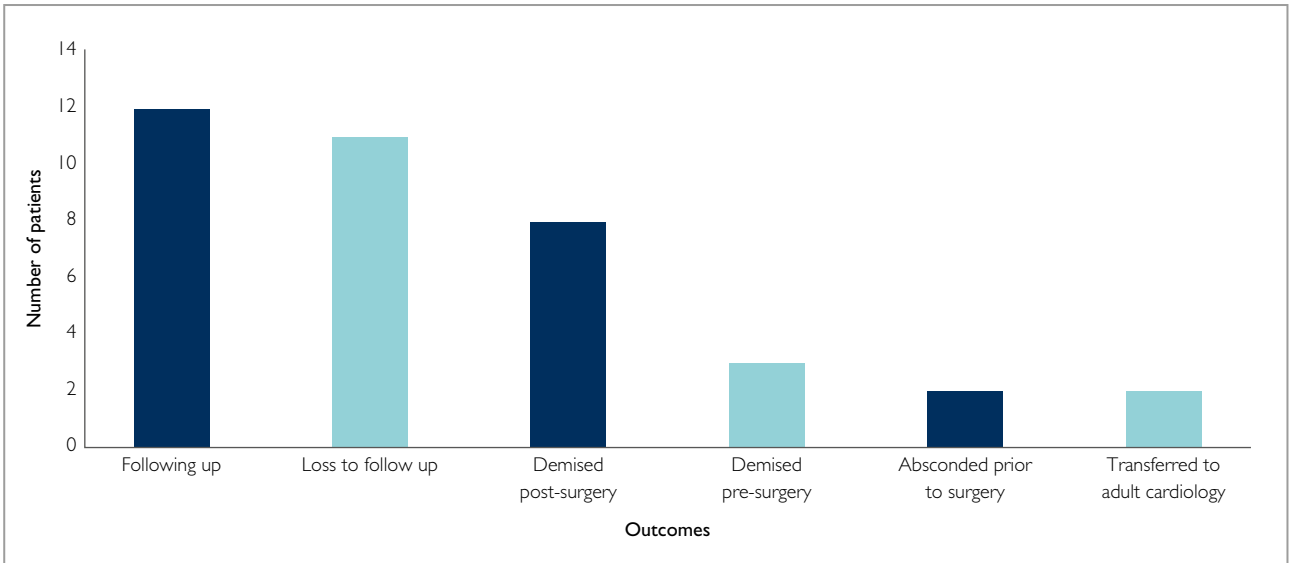

FIGURE 2: Illustrating the outcomes of the patients diagnosed with ALCAPA at the CHBAH from 1991 - 2018.

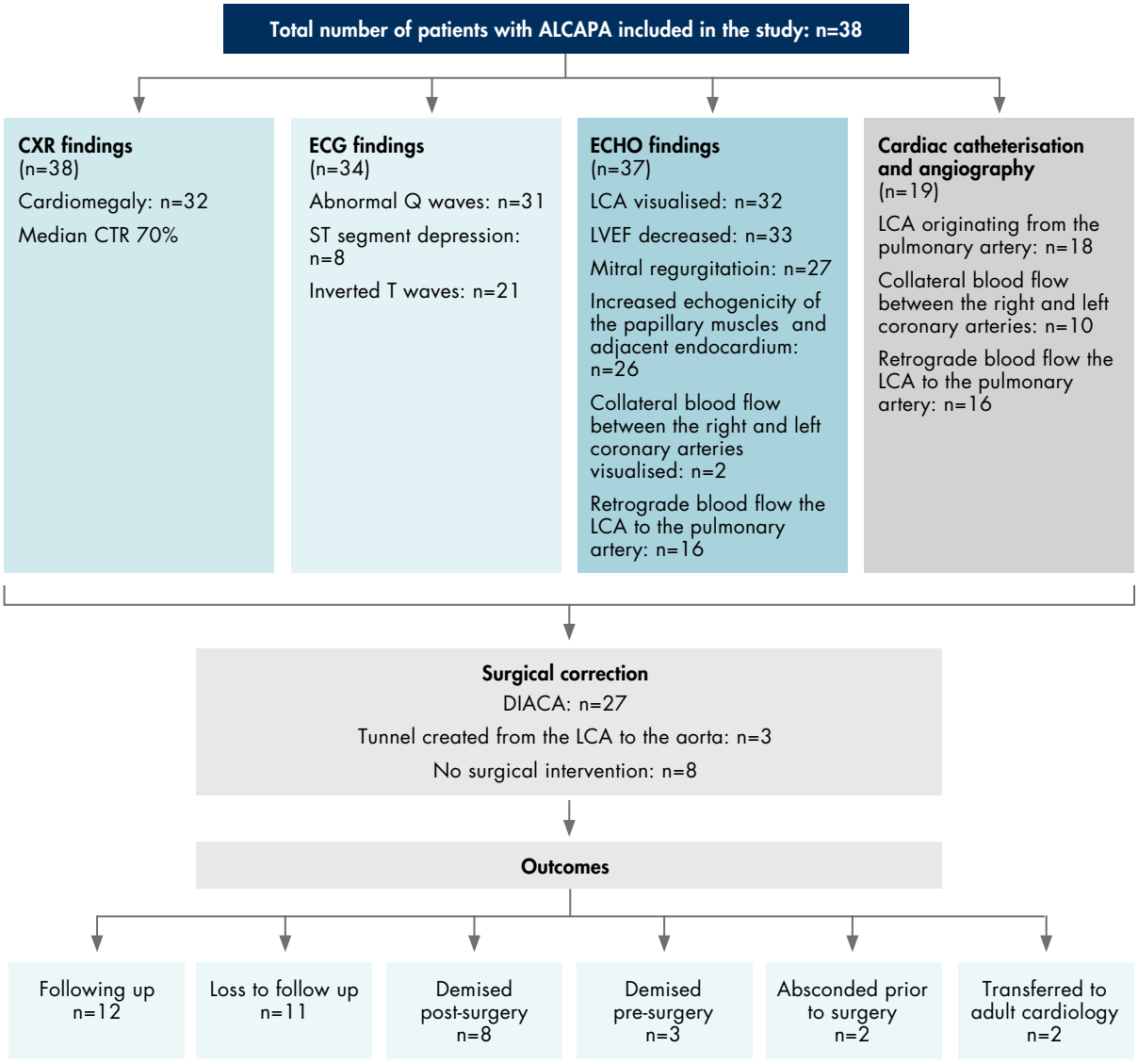

FIGURE 3: Schematic overview of the diagnostic findings and outcomes in the patients included.

DISCUSSION

ALCAPA was found to comprise 0.45% of all congenital cardiac conditions in our study, which is comparable with that of the global incidence. The median age of presentation of the study cohort was 4.3 months, which correlates with the drop in physiological hypertension of the newborn and the consequences thereof. The most frequent symptoms on initial presentation were dyspnoea, cough, and poor feeding, which is comparable with other studies.[1-4] The most common signs occurring in our study cohort at initial presentation were a displaced apex beat, respiratory distress, and hepatomegaly. An initial diagnosis of a lower respiratory tract infection was made in majority of the patients (71.1%).

Previous studies show that most patients have cardiomegaly present on CXR.[3,5] In our study cohort, 84.2% of patients were found to have cardiomegaly on CXR, with a median CTR of 70%. The presence of cardiomegaly is an important red alert to the possibility of an underlying cardiac disease and if it is not present, it may delay the diagnosis. Myocardial ischaemia ensues in the late neonatal and infant period when the pulmonary pressures decline, resulting in ischaemic changes which become evident on the ECG.[3] The majority of the patients, across all age groups (94.1% of the patients with ECGs) showed deep Q waves particularly in standard leads I and aVL which is a classical finding previously reported.[3] Other ischaemic changes such as T wave inversion and ST segment depression were also observed in the lateral and inferior leads.

TABLE VI: A univariate analysis for the variables listed and the LVEF at diagnosis.

Variable	Coefficient (95% Confidence Interval)	p-value
Age in months	0.20 (-0.62 - 0.47)	0.13
Gender	8.81 (-1.35 - 18.97)	0.09
Anthropometry		
Wasted	-12.98 (-32.00 - 6.04)	0.17
Severely wasted	-11.65 (-23.19 - 0.10)	0.05
CTR on CXR	-0.53 (-1.21 - 0.15)	0.12
Abnormal Q waves on ECG	-7.40 (-29.69 - 14.88)	0.50
ST depression on ECG	-1.84 (-14.27 - 10.60)	0.77
Inverted T waves on ECG	-1.32 (-12.18 - 9.53)	0.81
Mitral regurgitation on echocardiography	-5.30 (-17.10 - 6.51)	0.37
Retrograde blood flow from the LCA to the pulmonary artery on cardiac catheterisation and angiography	-4.19 (-30.50 - 22.12)	0.74
Presence of collateral blood flow from the right coronary artery to the LCA on cardiac catheterisation and angiography	-0.63 (-17.32 - 16.07)	0.94

Increased echogenicity of the anterolateral papillary muscle and the adjacent myocardium due to fibrosis and ischaemia, was present in 70% of our study patients. The median LVEF in the study cohort of patients was 34% which is lower compared to other studies.[1,15] This may be attributed to our study having more infant patients (81.6%) who are more likely to present with left ventricular dysfunction.[9,11] There was a high correlation between failure to thrive and wasting and a diminished LVEF. Poor feeding, as a result of heart failure, and a low LVEF could explain why there is a correlation between wasting and a low LVEF.

A study by Zheng, et al. found that an increased CTR, significant Q waves and T wave inversion, a lower LVEF, and more severe left ventricular dilatation was more prevalent in the infant group.[1] In our study, echocardiographic visualisation of the LCA originating from the pulmonary artery and the presence of left ventricular dilatation in the infant group were the only statistically significant findings when comparing the infant and non-infant groups. Older patients may have poorer echocardiographic acoustic windows that may mitigate against good visualisation of the LCA origin. In addition, older patients at presentation are likely to have developed good collateral blood flow and therefore may have less left ventricular dysfunction and dilatation.

Cardiac catheterisation and angiography confirmed the diagnosis of ALCAPA in several patients where the diagnosis was not clear on echocardiography in the study cohort. The confirmation of the diagnosis using echocardiography alone was possible in 86.5% of patients and improved to 94.7% when angiography was included.

An unusual case from the study cohort illustrating the diversity of the presenting symptoms was that of a patient with Trisomy 21 with a large PDA and severe reversible pulmonary hypertension, who was accepted for surgical ligation. During attempted surgical ligation of the PDA the patient became unstable haemodynamically. Eisenmenger's syndrome was suspected and surgery was abandoned. A subsequent cardiac catheterisation showed reversible pulmonary hypertension again, but a chance observation on review of the pulmonary angiogram showed the LCA originating from the main pulmonary artery. It is likely perfusion of the LCA was dependent on the high pulmonary pressures and following ligation of the PDA, the pulmonary pressures dropped, resulting in reduced perfusion of the LCA with ensuing myocardial ischaemia. The patient subsequently underwent a successful PDA ligation and DIACA of the ALCAPA.

The majority of patients in our study underwent surgical correction using the DIACA approach. Most of our patients showed normalisation of their reduced LVEF within 6 months. Similarly, mitral regurgitation improved from moderate and severe to mild mitral regurgitation with complete resolution at a median of 6 months post-surgery. The majority of patients were asymptomatic by the first follow up visit. An improvement in LVEF post-surgery does not preclude subclinical residual myocardial damage and perfusion deficits, as demonstrated by the late post-surgical MRI findings in a study by Alexi-Meskishvili,

TABLE VII: A Bivariate analysis of the demographic and diagnostic findings in the survival versus the non-survival groups.

Variable	Survival group Number of patients (% of the total number of patients for the variable stated)	Non-survival group Number of patients (% of the total number of patients for the variable stated)	p-value
Demographic	**n=14**	**n=11**	
Age			
Infant	11 (78.6)	11 (100.0)	0.23
Non-infant	3 (21.4)	0 (0.0)	
Gender			
Female	9 (64.3)	8 (72.7)	1.00
Male	5 (35.7)	3 (27.3)	
Anthropometry			
Normal	6 (42.9)	3 (27.3)	1.00
Wasting	5 (35.7)	4 (36.4)	
Severely wasted	1 (7.1)	1 (9.1)	
Radiographic findings	**n=14**	**n=11**	
Presence of cardiomegaly	13 (92.9)	9 (81.8)	0.57
ECG findings	**n=13**	**n=10**	
Rate			
Normal	11 (84.6)	8 (80.0)	1.00
Tachycardic	2 (15.4)	2 (20.0)	
Axis			
Normal	8 (61.5)	9 (90.0)	0.41
Right	2 (15.4)	0 (0.0)	
Left	3 (23.1)	1 (10.0)	
Abnormal Q waves	12 (92.3)	10 (100.0)	1.00
ST segment depression	2 (15.4)	3 (30.0)	0.62
Inverted T waves	9 (69.2)	6 (60.0)	0.69
Echocardiographic findings	**n=13**	**n=11**	
LCA visualised originating from the pulmonary artery	12 (92.3)	10 (90.9)	1.00
Decreased LVEF	13 (100.0)	9 (90.0)	0.44
Presence of left ventricular dilatation	9 (69.2)	7 (63.6)	1.00
Presence of mitral regurgitation	9 (69.2)	6 (54.5)	0.68
Dilated right coronary artery	6 (46.2)	5 (45.5)	1.00
Cardiac catheterisation and angiography	**n=6**	**n=4**	
Retrograde blood flow from the LCA to the pulmonary artery	5 (83.3)	3 (75.0)	1.00
Collateral blood flow from the right coronary artery to the LCA	3 (50.0)	2 (50.0)	1.00

et al.[16] Continued follow up post-surgical correction is therefore required.

In our study, the early mortality rate was 21.6% which is far higher than in other studies.[10,13,17] This may be attributed to various factors including a delay in the initial diagnosis, delays in transfers to a specialist cardiologist due to transport difficulties, delays in surgical correction due to long waiting lists or delays while attempting to optimise patients for open heart surgery.

Anomalous right coronary artery from the pulmonary artery (ARCAPA) is more rare than ALCAPA (incidence of 0.002%) and often an incidental finding.[18] Patients with ARCAPA may occasionally present with symptoms of myocardial ischaemia

when "coronary steal" occurs from the LCA into the right coronary artery.[18] The study cohort included a male child with ARCAPA who presented at 9 months with failure to thrive, dyspnoea, and poor feeding. The CXR showed cardiomegaly and the ECG demonstrated poor right ventricular forces. Echocardiography showed a possible ARCAPA, a decreased LVEF of 46%, and moderate mitral regurgitation. The diagnosis was confirmed on cardiac catheterisation and angiography and the patient underwent surgical reimplantation of the right coronary artery onto the aorta. His left ventricular function normalised 67.4 months after surgery which is considerably longer than the group with ALCAPA.

Our study showed no statistical difference in the diagnosis and outcomes of patients between the first and second decades of the study period, suggesting that despite improvements in diagnostic technologies, the diagnosis of ALCAPA depends more on the awareness of ALCAPA as a possible diagnosis. An early diagnosis can allow for rapid and appropriate surgery and improved patient outcomes.

Limitations

A small sample size was a major limitation of our study; however, similar sample sizes were documented in other studies mainly due to the rarity of this pathology. The retrospective nature of the study contributed to the absence of important data in some patients. In addition, there was a major loss to follow up of patients, which placed a limitation on the post-surgical outcome and survival analysis. The cause of poor follow up is multifactorial in our setting and may be attributed to transport difficulties created by financial constraints, patient demise or patients returning to other provinces or countries of origin.

CONCLUSION

ALCAPA is a rare congenital cardiac condition resulting in left ventricular dysfunction and it is amenable to surgical intervention with good outcomes. Due to the non-specific presenting symptoms and clinical manifestations, a high index of suspicion by clinicians is crucial in prompting further investigations such as an ECG which has features commonly occurring in patients with ALCAPA. An awareness of the classical ECG features by medical staff referring patients from secondary hospitals would contribute to making a diagnosis and then promptly referring the patients to a tertiary centre for confirmation of the diagnosis and management.

Conflict of interest: none declared.

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