

## Case Report

# Bland-White-Garland syndrome in a neonate with review of literature

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## ABSTRACT

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) or the Bland-White-Garland syndrome was first described in the year 1866 and postulated in 1933. It is an intriguing entity in the field of paediatrics as well as paediatric cardiology due to its varied presentation in the paediatric age group. This congenital heart anomaly is reported in less than 0.5% of all the congenital heart diseases. In the past, infant deaths were frequent due to not diagnosing it promptly. Presently, the prognosis for these patients with ALCAPA has dramatically improved as a result of the scientific advancements in the field of cardiology such as early diagnosis using echocardiography and improvements in cardiovascular surgical techniques. Interestingly many treating doctors are not aware about the significance of the ALCAPA and can be missed easily. The chance of ALCAPA should be searched in every infant or children with non specific symptoms of incessant crying and feeding difficulty to achieve better outcome and to prevent its complication such as sudden death. This case study reports ALCAPA in a 23 day old neonate.

**Key words:** ALCAPA, Bland-White-Garland syndrome, Crying newborn, congenital heart diseases, Coronary steal, Myocardial ischemia, Antero-lateral myocardial infarction

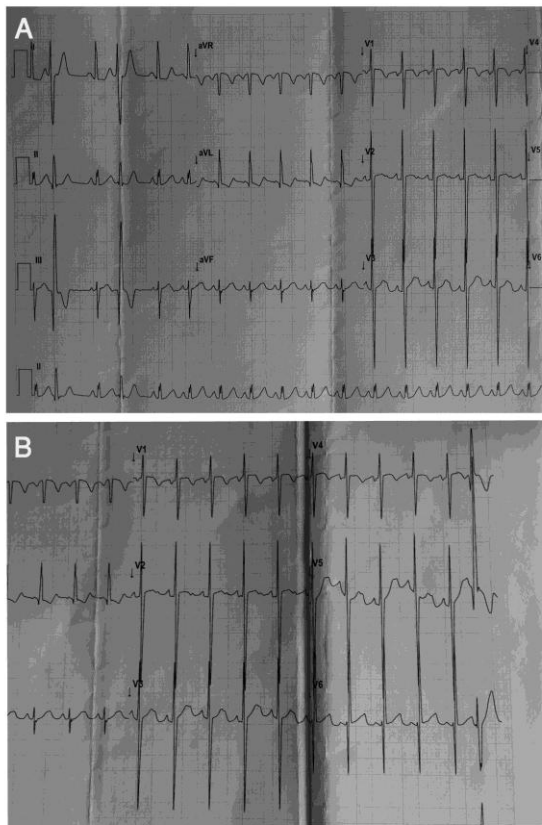
## INTRODUCTION

Bland-White-Garland syndrome, a rare paediatric cardiac anomaly is the ALCAPA syndrome (anomalous origin of the left coronary artery from the pulmonary artery), in which the left coronary artery has an unexpected anomalous origin from the pulmonary artery [1]. The significance of diagnosing ALCAPA lies in the fact that it is associated with serious complications such as myocardial ischemia and infarction in early infancy, congestive heart failure, and sometimes death during the early infantile period if appropriate treatment is delayed [1,2]. The babies experience angina pain due to the myocardial ischemia but due importance is often not given as they present with symptoms of pain or distress (often mistaken for colic), irritability, rapid breathing sweating and poor feeding. Early diagnosis, especially electrocardiogram will reveal the classic changes of ALCAPA and prompt surgical treatment of the disease can be life saving. The development of the collateral circulation in ALCAPA also decides the variation in clinical presentations and outcome. Literature review has reported that many have died in the infantile period and also in the young age [2]. This article reports a case of ALCAPA in a 23 day old neonate.

## CASE REPORT

A 23 day old male baby was seen in the outpatient with breathing difficulty since the previous night. He was treated for symptoms of nasal block and low grade fever for the last 3 days. He was breathing spontaneously with no cyanosis. He was exclusively breastfed and there was no history of any aspiration of breast milk or oral secretions. Birth history revealed he was born to non-consanguineous parents of 24 year old mother at 38 weeks of gestation via spontaneous vaginal delivery. Birth weight was 3.1 kg with normal apgar score. On general physical examination, baby was tired, crying and sweating with a grunt. He was febrile (100°F), heart rate 142/minute, respiratory rate 66/minute, weight 3.36 kg. anterior fontanelle soft and flat, skin was pale. His oxygen saturation was 96%. All peripheral pulses were equally felt. Blood pressure was 84/56 mm Hg in right upper limb, 83/56 mm Hg in left upper limb, 84/56 mm Hg in right lower limb and 86/57 mm Hg in left lower limb. His respiratory effort remained good and was very irritable. The breath sounds were equal on both sides of thorax with increased conducted sounds. Cardiovascular system examination revealed S1, S2 normal, a 2/6 systolic murmur over the precordium with cardiomegaly. There was no hepatomegaly. A diagnosis of congenital heart disease (CHD) was considered and the infant was placed under humidified oxygen. His general condition improved

with symptomatic treatment given for fever with paracetamol drops but was crying and had episodes of becoming pale. His chest radiograph (CXRAY) showed cardiomegaly with prominent bronchovascular markings. The electrocardiogram (ECG) showed sinus tachycardia with occasional ectopic premature complexes, left ventricular hypertrophy and typically, an anterolateral infarction pattern with small q, T inversion and depression of ST-segment in anterolateral leads. q waves were absent in leads III, and aVF and were deep (>5mm) in V<sub>5</sub>, and V<sub>6</sub> (Figure 1A and B).



**Figure 1**

ECG prompted us to aggressively think of the possibility of ALCAPA. 2D Echocardiogram showed a dilated left atrium and left ventricle, posterior and lateral wall akinetic and also scarred associated with mitral regurgitation (MR). The laboratory investigations were done: A quick glucose check was 175 mg/dl. Hemogram revealed Hb (12 g/dl), PCV (33.7%), total count (10000/ $\mu$ l), neutrophils (47%), lymphocytes (48%), eosinophils (2.3%), monocytes (2.5%), basophils (0.2%), ESR (31mm/hr), and platelets count (210000/ $\mu$ l) were all found to be

normal. Serum Na<sup>+</sup> 133 meq/l, K<sup>+</sup> 4.4 meq/l, Cl<sup>-</sup> 91 meq/l, bicarbonate 26 meq/l, glucose 144 mg/dl, total Ca<sup>++</sup> 9.0 mg/dl. C Reactive protein was normal (<0.6 mg/dl). The baby was promptly referred to pediatric cardiothoracic centre. These non invasive interventions (ECG, CXRAY, 2DECHO) along with the clinical presentations motivated to diagnose ALCAPA and referred to Paediatric Cardiothoracic centre.

## DISCUSSION

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a cardiac anomaly, when the left coronary artery (LCA) is connected to the pulmonary artery (PA) instead of to aorta. Bland et al. (1933) studied this pathology in autopsy findings, correlated them as Bland-White-Garland syndrome [3]. There is no male or no racial predilection, as in our report and frequency is equal in both sexes. There is no mode of inheritance and ALCAPA is not associated with any syndromes or genetic noncardiac conditions [4].

ALCAPA is congenital. Myocardium is deprived of oxygen due to the anomalous origin as well as due to "coronary steal". Before birth, high PA pressures and normal antegrade flow in the LCA ensures adequate blood supply to the left ventricle (LV) myocardium and ALCAPA is asymptomatic. After birth, in the neonatal period the low blood pressure in the PA causes blood from the abnormal LCA to flow toward the PA instead of toward the heart (coronary steal). Further, damages the myocardium in babies with ALCAPA. So the collateral circulation which develops between the right and left coronary artery (RCA and LCA) during closure of the ductus arteriosus and a fall in the pulmonary pressure will determine the extent of myocardial ischaemia [5]. Furthermore, coronary steal develops over time in babies with ALCAPA, if the condition is not treated early. Coronary steal condition causes underperfusion of LV myocardium, CHF develops as a result of LV dysfunction and the mitral valve insufficiency [5].

Myocardial ischemia causes significant chest pain and the baby starts having crying episodes which will increase when the infant is feeding and crying. This eventually increase the myocardial oxygen consumption leading to infarction of the anterolateral LV free wall and causing mitral valve papillary muscle dysfunction and mitral insufficiency. These babies have pallor, irritability, and diaphoresis after feeding, which are mistaken as colic. Features of CHF such as tachypnea, tachycardia, diaphoresis, poor feeding, leading to poor weight gain are seen in about 85% of

patients present within the first 1-2 months of life [1]. The delayed presentations of myocardial ischemia, with MR later in childhood or even adulthood with periodic dyspnea, angina pectoris, syncope, or sudden death reported [1,6]. A case of 72 year old woman with ALCAPA has been reported [7].

A thorough clinical examination is necessary and usually reveals a systolic murmur of MR and heard well in the apical left precordial region in 70% of cases as in our case [1]. Signs of CHF may be present. There is prominence of the LV precordial impulse which is displaced both inferiorly and laterally. Also increased intensity of the pulmonic component of the second heart sound may be seen with PA hypertension secondary to elevated left atrial pressure. Hepatic enlargement may be observed, and the peripheral pulses may be diminished in intensity secondary to low cardiac output in cases of severe CHF. In such infant, a possibility of conditions such as dilated cardiomyopathy, coronary artery fistula, mitral valve insufficiency, viral myocarditis, large left to right shunts and mixing lesions with increased pulmonary blood flow should be considered in the differential diagnosis [1,8].

The diagnosis of ALCAPA is based on high degree of clinical suspicion but the important tool is the ECG [9]. In ALCAPA, an anterolateral infarct pattern with wide (>30 msec) q waves is observed in leads I, aVL, absent q waves in leads II, III, and aVF. Poor R wave progression across the precordial leads, with sudden shift to qr is also an important finding to clinch the diagnosis. CXRAY reveals the cardiomegaly, with or without pulmonary venous congestion [8,9]. Cardiac isoenzymes assay are not helpful in diagnosis. 2DECHO with Doppler color flow mapping is often diagnostic. The use of color flow velocity mapping is helpful in demonstrating retrograde flow from the anomalous left coronary into the pulmonary trunk [9]. An increased echogenicity of left ventricular papillary muscles, scarring in the myocardium can be seen along with MR, left ventricular dysfunction along with abnormalities of cardiac wall motion [9]. Aortography or selective right coronary arteriography is a standard diagnostic tool [10].

The treatment of ALCAPA involves initially stabilization with control of CHF by using diuretics, after load reduction medications, and inotropic drugs. Oxygen reduces pulmonary vascular resistance and magnify coronary steal from the RCA into the PAs and so administration of 100% oxygen is postulated to be deleterious. Surgical intervention is the procedure of

choice as the treatment with afterload reduction and use of inotropic drugs again may worsen ischemia [11]. Ligation of the LCA at its origin from the main PA was performed in the past and the patient remained at risk for ischemic episodes and sudden death. Current revascularization procedures ensures creating a 2, coronary artery system via either direct reimplantation, an anastomosis of left subclavian artery to coronary artery, a saphenous vein bypass graft or the well known Takeuchi procedure which is the creation of an aortopulmonary window and an intrapulmonary tunnel extending from the anomalous ostium to the window [12]. Mitral valve reconstruction is not necessary. Most patients do extremely well and CHF symptoms resolve along with normalization of ECG changes. Diuretics and afterload reduction have to be continued till significant improvement of left ventricular function and reduction of MR. Complications are rare after surgery. Cardiac dysrhythmias should be aggressively monitored. Cardiovascular magnetic resonance (CMR) is noninvasive tool, without ionizing radiation tool to look for anterolateral subendocardial myocardial fibrosis after the surgery. Stress adenosine CMR perfusion detects reversible ischemia in these patients due to occlusion of LCA [13].

This case study concludes that awareness of ALCAPA among the treating physicians is very important for not missing this serious life saving condition in a crying infant or in a child with a dilated LV. Surgical revascularisation procedures are lifesaving and is the treatment of choice. ALCAPA has good prognosis, if diagnosed early with the simple non invasive tools such as ECG and a diagnostic 2DECHO to confirm it.

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