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"ANOMALIES ORIGIN OF LEFT CORONARY ARTERY" ITS EMBRYOLOGICAL BASIS AND CLINICAL SIGNIFICANCE

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ABSTRACT

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The Coronary artery development is a delicate process that includes a complex interaction among the pericardium and the myocardium which leads to involving of multiple cellular pathways. There has been a growing engrossment in the study of the coronary artery development. It helps to identify some of the important causes of anomalous coronary development. There are different kinds of anomalous coronary arteries, having variable risk in myocardial ischemia, malignant arrhythmias, and sudden cardiac death also. A good understanding of this process could help us insharing new avenues in the treatment of clinically relevant anomalous coronary arteries.

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Introduction

In this anomaly, the left coronary artery from the pulmonary artery is typically a rare heart defect. The left coronary artery that carries blood into the heart muscle, and it is connected to the pulmonary artery instead of connecting to the aorta. This condition is one which causes the poor cardiac function in infancy or child. Anomalous left coronary artery from the pulmonary artery otherwise shortly known as (ALCAPA) it is present at the birth, which means the congenital. [1]ALCAPA was firstly mentioned in 1866 and the first clinical study in conjunction with autopsy findings was described by Bland and colleagues in 1933, so this anomaly is also named as Bland-White-Garland syndrome. This anomaly may result in two forms [1, 2]. The abnormal septation of the conotruncus into the aorta and pulmonary artery, or the persistence of the pulmonary buds together with involution of the aortic buds that finally leads to form the coronary arteries. ALCAPA is usually an isolated cardiac anomaly but in certain rare incidences, it has been described with patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot and coarctation of the aorta [2,3].

During the fetus-developing period, the pulmonary artery pressure is equal to the systemic pressure that allows satisfactory myocardial perfusion from the pulmonary artery through the anomalous coronary artery [2-5]. But after birth, the pulmonary artery contains desaturated blood pressure that rapidly falls below systemic pressure. Due to this, the left ventricle with its huge need for oxygen is perfused with desaturated blood at low pressure. Early diagnosis and prompt surgical treatment of the disease can save the life of the victim. Survival to adulthood is quite uncommon. Infants experience myocardial infarction and congestive heart failure, and approximately 90% die within the first year of life [3,4,5]. Once the patient has been stabilized with the condition, different surgical approaches can be proposed to the patient.

Simple ligation is used to prevent a steal from the myocardium and it is associated with the significant short- and long-term complications. This is because, after ligation, the heart is converted to the one-vessel coronary system, making it entirely dependent upon right coronary artery[5,6]. This study material aims to through insight knowledge about the left coronary artery from the pulmonary artery or Bland-White-Garland syndrome. This helps to understand their association with the left coronary artery and various descriptions regarding its development and complications [6]. Incidence

The Anomalous origin of the left coronary artery arises from the pulmonary artery (ALCAPA). This is a rare congenital cardiac anomaly in which medical council recorded that approximately 0.25-0.5% of all is congenital heart disease. The incidence of left coronary artery does not vary geographically. This is not considered as an inheritable congenital cardiac defect [5,6].

Ontogenesis of normal development of coronary vessels

Before 1989, Development of coronary arteries was generally believed that coronary arteries and veins were developed from the outgrowths from the aorta and the systemic venous sinus respectively [6,7]. But in a study lead by Bogers et al revealed that the coronary arteries were identified even before the presence of coronary arterial orifices, which led to the a new concept of ingrowth instead of outgrowth of the coronary arterial vasculature. Epicardium is essential for coronary vessel development [6]. The term epimyocardium has been used in implying the epicardium and myocardium may have a common origin. The epicardium originates from an extra cardiac structure and the proepicardium and which is located near to the venous pole of the heart just above to the liver primordium. Epicardial pole of the heart lies just above to the liver primordium.

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Epicardial cells form a sheet of single cells which covers the heart and are necessary for coronary vessel formation [7,8]. An elaborate interaction of epicardial cells with myocardial a cells that lead to the transformation of the epicardial cells into a mesenchymal cells, as precursors of building blocks of coronary vessels. The hematopoietic progenitor cells also play an important role in the transformation of epicardial into mesenchymal cells .In humans, at about the 25thembryonic day, the vessel-like structures that are observed between in the space of the epicardium and the myocardium [9]. Then it should be emphasized that at this point there is no blood supply in these vessel-like structures and which are not all contiguous.[10,11]. In a matter of days these structures fuses to form a newly vascular plexus. This diffused elaborate vascular network ultimately get contacts and will penetrates with the aortic root and leads to abrupt exposure to the systemic pressure and high flow. These changes result in the maturation of these vessels, including the migration of smooth-muscle cells and the proper arrangement of these cells and further growth of some vessels, and regression of others through apoptosis. Variations in this delicate controlled process may leads to the congenital coronary artery anomalies [12,13].

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Fig 1: Schematic representation shows the ontogenesis for the normal development of Coronary arteries.

Ontogenesis for the anomalous origin of left coronary artery

The coronary arterial anomalies is classified into 2 types of anatomic subsets: with the origin of the anomalous coronary artery from the opposite aortic sinus, and with the origin of the anomalous coronary artery from the pulmonary artery (PA)[14,15]. The ALCAPA anomaly may result from (i) abnormal septation of the conotruncus into the aorta and pulmonary artery, or from (ii) persistence of the pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries. The Anomalous coronary artery from the opposite sinus includes the left coronary artery (LCA) arising from the right sinus. If the anomalous coronary artery courses in the sulcus between the PA and the aorta, which occurs in the ALM, were there is a high risk of malignant arrhythmias and sudden death of the patient [16,17]. The increased risk may be due to the intra-arterial pump of the anomalous coronary which results in the compression or the

larger myocardial mass is supplied by the anomalies of this pattern.

This presence of the slit-like origin and the intramural course of the proximal coronary artery that emerges from the contralateral sinus probably plays a major role in coronary ischemia and malignant arrhythmias. A coronary artery originating from the PA is much more rare, and occurring in about 1 in 300000 medical cases, and more commonly involving in the LCA emerging from the main Pulmonary Artery.[18].



Fig-2: Schematic representation shows the abnormal origin of left Coronary artery from the pulmonary trunk Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but serious congenital cardiac anomaly. ALCAPA was first described in 1866. The first clinical description in conjunction with autopsy findings was described by Bland and colleagues in 1933, so the anomaly is also called Bland-White-Garland syndrome [19,20]. In 1962, Fontana and Edwards reported a series of 58 postmortem specimens that demonstrated that most patients had died at a young age.

Presently, the prognosis for patients with ALCAPA is dramatically improved as a result of both early diagnosis using echocardiography with color flow mapping and improvements in surgical techniques, including myocardial preservation [20]. ALCAPA is usually an isolated cardiac anomaly but, in rare incidences, has been described with patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot, and coarctation of the aorta. Extremely rare variations of anomalous origin of the coronary arteries from the main pulmonary artery includes, (i) the left anterior descending or circumflex branches, or (ii) The right coronary, often discovered as an incidental finding on autopsy or (iii) Both the right and left coronary arteries, a circumstance not compatible with survival[21].

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) does not present prenatally because of the favorable fetal physiology that includes equivalent pressures in the main pulmonary artery and aorta secondary to a nonrestrictive patent ductus arteriosus, and relatively similar oxygen concentrations due to parallel circulations [22]. This results in normal myocardial perfusion and, therefore, no stimulus for collateral vessel formation between the right and left coronary artery systems is present.

Shortly after birth, as the circulation becomes one in series, pulmonary artery pressure and resistance decrease, as does oxygen content of pulmonary blood flow. This results in the left ventricular myocardium being perfused by relatively desaturated blood under low pressure, leading to myocardial ischemia; low pressure is more important in causing decreased myocardial perfusion [22].

Initially, myocardial ischemia is transient, occurring during periods of increased myocardial demands, such as when the infant is feeding and crying. Further increases in myocardial oxygen consumption lead to infarction of the anterolateral left ventricular free wall. This often causes mitral valve papillary muscle dysfunction and variable degrees of mitral insufficiency [23].

Collateral circulation between the right and left coronary systems ensues. Left coronary artery flow reverses and enters the pulmonic trunk due to the low pulmonary vascular resistance (coronary steal phenomena) [23, 24]. As a result, left ventricular myocardium remains under perfused. Consequently, the combination of left ventricular dysfunction and significant mitral valve insufficiency leads to congestive heart failure (CHF) symptoms (e.g., tachypnea, poor feeding, irritability, and diaphoresis) in the young infant. Inadequate myocardial perfusion likely causes significant chest pain and these symptoms of myocardial ischemia may be misinterpreted as routine infantile colic [25].

The main aim of this study is to give a major overview of normal coronary anatomy, which describes the common variants of the normal and the summarized part of typical patterns of the anomalous coronary arteries anatomy. Previously these anomalies where basically could only be described from the autopsy reports due to the huge developments in the imaging modalities in the recent years [26]. These anomalies can now be detected through noninvasively method. The use of the computed tomography (CT) angiography and the cardiac magnetic resonance (CMR) to define these anomalies gives scope for the physicians to perform better treatment for the patient. This study will be heavily supported with the imaging taken from the invasive coronary angiography, CT and the CMR which helps to illustrate the nature of the anomaly that is possible, thus we can treat the patient through various surgical process and proper medications [27, 28].

Conclusion

The left coronary artery which is arising from the pulmonary artery is typically a rare heart defect. The left coronary artery that carries blood into the heart muscle, and it is connected to the pulmonary artery instead of connecting to the aorta. This condition is one which causes the poor cardiac function in infancy or child. Anomalous left coronary artery from the pulmonary artery otherwise shortly known as (ALCAPA) it is present at the birth, which means the congenital. In the normal heart, the left coronary artery starts in the aorta. The aorta is the major blood vessel that takes oxygen-rich blood from the heart to the rest of the body.

In children with ALCAPA, the left coronary artery starts at the pulmonary artery. The pulmonary artery is the major blood vessel that takes oxygen-poor blood from the heart to the lungs. When this defect occurs, blood that is lacking in oxygen is carried to the left side of the heart. Therefore, the heart does not get enough oxygen. When the heart muscle is deprived of oxygen, the tissue begins to die. This condition leads to a heart attack in the baby.

A condition known as "coronary steal" further damages the heart in babies with ALCAPA. The low blood pressure in the pulmonary artery causes blood from the abnormal left coronary artery to flow toward the pulmonary artery instead of toward the heart. This results in less blood and oxygen to the heart. This problem will also lead to a heart attack in a baby. Coronary steal develops over time in babies with ALCAPA if the condition is not treated early [27,28].

During the fetus-developing period, the pulmonary artery pressure is equal to the systemic pressure that allows a satisfactory myocardial perfusion from the pulmonary artery through the anomalous coronary artery. But after birth, the pulmonary artery contains desaturated blood pressure that rapidly falls below systemic pressure. Due to this, the left ventricle with its huge need for oxygen is perfused with desaturated blood at low pressure. Early diagnosis and prompt surgical treatment of the disease can save the life of the victim. Survival to adulthood is quite uncommon. Babies experience myocardial infarction and congestive heart failure, and approximately 90% die within the first year of life. These anomalies can now be detected through non-invasively method. The use of the computed tomography (CT) angiography and the cardiac magnetic resonance (CMR) to define these anomalies gives scope for the physicians to perform better treatment for the patient [29].

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