ORIGINAL ARTICLE

Prevalence of Coronary Artery Anomalies and variants in Jinnah Hospital

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ABSTRACT

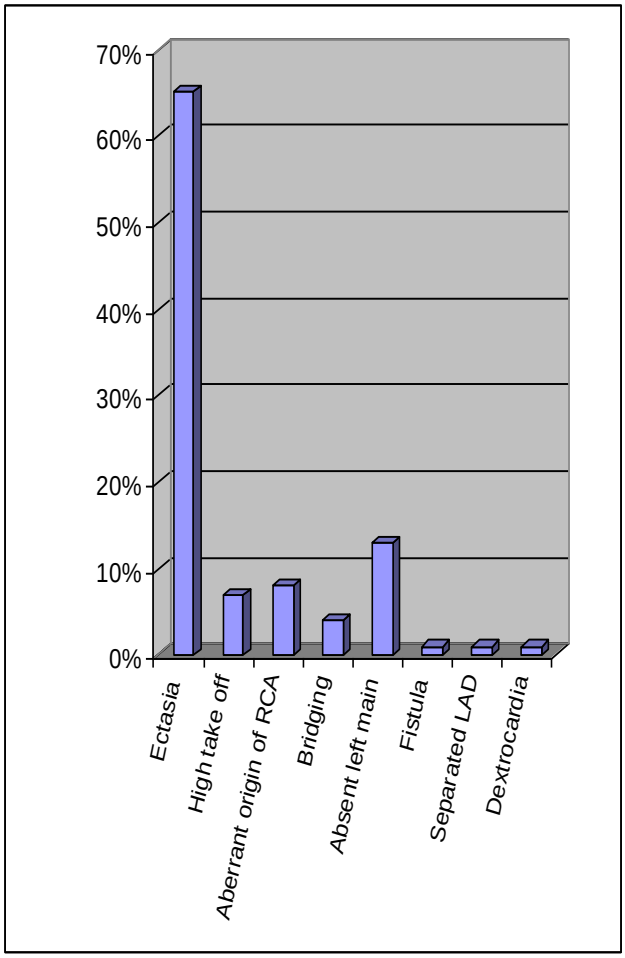
Aim: To assess the prevalence of variants and anomalies of the coronary artery tree in patients who underwent coronary angiography for suspected or known coronary artery disease
Method: The diagnostic angiographic reports of the angiograms done in the department of cardiology Jinnah Hospital Lahore in 2011 were reviewed. In those in which a congenital anomaly was diagnosed, the angiography film was studied.
Results: Nine hundred fifty six reports were reviewed describing 26 patients with 26 anomalies with a prevalence of about 2.7% including: anomalous origin of the right coronary artery, coronary artery fistulae, separate origin of anterior descending artery, absent Left Main, Bridging and others. Angiographic studies were done because of: angina (65%), dyspnea (25%) and atypical chest pain (10%). Adult congenital anomalies of the coronary arteries are not very common and are usually casual findings of diagnostic angiographic studies. Ectesia, absent Left main and Aberrant Right coronary artery origin were the most frequently diagnosed in our population.
Keywords: Coronary artery, anomalies, prevalence

INTRODUCTION

Anomalies of the coronary arteries may be found incidentally in 0.3%–1% of healthy individuals [01].Although coronary artery anomalies are far less common than acquired coronary artery disease, their impact on premature cardiac morbidity and mortality among young adults is not trivial. In a study by Eckart et al [02] of 126 nontraumatic sudden deaths in young adults, cardiac abnormality was found in 64 cases (51%), with coronary artery abnormalities being the most common cardiac abnormality (39 of 64 patients [61%])

PATIENTS AND METHODS

A total of 956 patients (589 male, mean age 50.5 ± 10.9) were reviewed for coronary artery variants and anomalies. The coronary dominance pattern results were: right 91%; left 5.8%; balanced 2%. The intermediate branch (means trifurcation) was present in the 4%. A variable number of diagonals (one, 21%; two, 59.7%; more than two, 24%; none, 1.3%) and marginal (one, 45.2%; two, 46.2%; more than two, 18%; none, 0.6%) were visualized. Single or associated coronary anomalies occurred in 9% of the patients, with the following distribution: 26 anomalies of origin and course, 59intrinsic anomalies (03 myocardial bridging, 56 aneurisms), 01 fistulas.
coronary anomalies



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Coronary anomalies:

ANOMALIES OF ORIGIN
High take off
Multiple Ostia
Single coronary artery
Anomalous origin of coronary artery from pulmonary artery
Origin of coronary artery or branch from opposite or non-coronary sinus and an anomalous retroaortic, interarterial, prepulmonic, septal course
Anomalies of course/intrinsic abnormalities
Myocardial Bridging
Duplication of Arteries
Ectesia
Anomalies of termination
Coronary Artey fistula
Coronary Arcade
Extracardiac termination

Modified from Angelini P. Normal and anomalous coronary arteries: definitions and classification. *Am Heart J.* 1989; 117:418– 434.

DISCUSSION

Angelini and coworkers[3] propose that, because of its substantial variability, normal and anomalous coronary anatomy should be characterized. Accordingly, an anomaly should be defined as any coronary pattern with a feature (number of Ostia, proximal course, termination, etc) "rarely" encountered in the general population.

We favor the following definitions: *normal*, any morphological feature observed in >1% of an unselected population; *normal variant*, an alternative, relatively unusual, morphological feature seen in >1% of the same population; and *anomaly*, a morphological feature seen in <1% of that population. (Van Camp and coworkers[4] reported that coronary anomalies cause 11.8% of deaths in US high school and college athletes. According to the Sudden Death Committee of the American Heart Association[5], coronary anomalies cause 19% of deaths in athletes. Moreover, Burke and colleagues[6] reported that in 14- to 40-year-old individuals, coronary anomalies are involved in 12% of sports-related sudden cardiac deaths versus 1.2% of non-sports-related deaths. In assessing 162 sudden deaths in a young general population, Drory and associates[7] found only 1 coronary anomaly. Similar findings suggest that coronary anomalies can be lethal only during or shortly after strenuous physical activity, typically in young individuals[7,8,9].

Coronary anomalies might have clinical consequences other than those strictly related to myocardial ischemia ; these consequences might include volume overload (in cases of coronary fistulas), aortic-root distortion (in cases of very large coronary fistulas or aneurysms), bacterial endocarditis, complications during aortic valve surgery or coronary angioplasty, and misdiagnosis (as in many cases of "missing" coronary arteries)[3]. However, because the coronary vessels primarily supply metabolic support to the dependent myocardium, physiological alterations in this function should be the main consideration. Unlike effort-related ischemia typical of fixed obstructive lesions, ischemia associated with coronary anomalies is reproducible with stress testing or is able to be fixed in only a few conditions (ALCAPA, coronary stenosis, or coronary atresia). In other anomalies, ischemia occurs only under inconsistent or extreme clinical conditions[8,9,14,15,35,36].

High takeoff" refers to the origin of either the RCA or the LCA at a point above the junctional zone between its sinus and the tubular part of the ascending aorta. Vlodaver et al[10] reported that both coronary ostia were situated above the sinotubular junction in 6% of randomly selected adult hearts. High take-off of the coronary arteries usually presents no major clinical problems, but it may cause difficulty in cannulating the vessels during coronary arteriography. Selective intubation of the coronary artery may be extremely difficult, especially when the RCA is anomalously located high over the left coronary sinus[11]. In our study it was found to be around 0.6%

In multiple ostia, typically either the RCA and the conus branch arise separately, or the LAD and LCx arteries arise separately with no LCA. An aberrant conus artery arising separately from the RCA is particularly at risk for injury from ventricu-lostomy or other maneuvers performed during heart surgery[12]. Separate ostia of the LCA and LCx artery may occur in a small percentage (0.41%) of individuals with otherwise normal anatomy[13]. Although multiple ostia represent a technical difficulty for the angiographer, they may also allow alternate collateral sources in patients with proximal coronary artery disease[14]. In our study the Left main was absent in about 1.1% of patients leading to separate origin of LCx and RCA.

The four recognized patterns of an anomalous origin of a coronary artery from the opposite or noncoronary sinus are *(a)* the RCA arising from the left coronary sinus, *(b)* the LCA arising from the right coronary sinus, *(c)* the LCx or LAD artery arising from the right coronary sinus, and *(d)* the LCA or RCA (or a branch of either artery) arising from the noncoronary sinus. In these anomalies, the coronary ostium may be at the normal level, or the involved artery may have a high or low takeoff[14]. Moreover, a coronary artery arising from the opposite or noncoronary sinus can take any of four common courses, depending on the anatomic relationship of the anomalous vessel to the aorta and the pulmonary

trunk: *(a)* interarterial (i.e., between the aorta and the pulmonary artery), *(b)* retroaortic, *(c)* prepulmonic, or *(d)* septal (subpulmonic)[15]. It is of great clinical importance which course is taken. Although retroaortic, prepulmonic, and septal (subpulmonic) courses seem to be benign, an interarterial course carries a high risk for sudden cardiac death[12,16,17].

The LCA arises from the right sinus of Valsalva as a separate vessel or as a branch of a single coronary artery in 0.09%–0.11% of patients who undergo angiography[18]. An interarterial course may be seen in up to 75% of patients with this anomaly[18, 19], who are at high risk for sudden cardiac death due to the acute angle of the ostium, the stretch of the intramural segment, and the compression between the commissure of the right and left coronary cusps. However, this anomalous LCA may also take a retroaortic, prepulmonic, or septal (subpulmonic) course[20]. Either the LCx artery or the LAD artery may anomalously arise from the right sinus of Valsalva. The LCx artery is the artery that most commonly arises from a separate ostium within the right sinus or as a proximal branch of the RCA (approximately 0.32%–0.67% of the population)[20]. Several reports have shown that this anomalous LCx artery passes behind the aortic root[19,21,22,23]; fortunately, this anomaly has not been associated with death. The LAD artery may arise from the right sinus in tetralogy of Fallot, double outlet right ventricle, and transposition complexes, but rarely in patients with otherwise normal hearts. It may take either an interarterial or a prepulmonic course[14]. Although either the RCA or the LCA may arise from the noncoronary sinus, both of these anomalies are rare in an otherwise normal heart[24,25] and usually have no clinical relevance. These anomalies may also be seen with transposition of the great arteries [14].In our study the aberrant origin of the RCA from the LCC was the only origin abnormality found with all having a retroaortic course with a percentage of 0.7%

Myocardial bridging is caused by a band of myocardial muscle overlying a segment of a coronary artery. It is most commonly localized in the middle segment of the LAD artery[26,27]. There is some discrepancy between the prevalence of myocardial bridging at angiography (0.5%–2.5%) and that at pathologic analysis (15%–85%)[27]. The cause for this discrepancy is presumed to be the fact that myocardial bridging often occurs without overt symptoms, so that patients are rarely referred for coronary angiography[28]. In some cases, however, myocardial bridging is responsible for angina pectoris, myocardial infarction, life-threatening arrhythmias, or even death[26]. The standard of reference for diagnosing myocardial bridges is coronary angiography, at which a typical “milking” effect and a “step down–step up” phenomenon induced by systolic compression of the tunneled segment may be seen[28]. In contrast, multi–detector row CT clearly shows the intramyocardial location of the involved coronary arterial segment[27]. The ECG-gated reconstruction window used in standard multi–detector row CT of the coronary artery is usually positioned within the diastolic phase for maximal vasodilatation and minimal motion artifacts[29]. However, when there is suspicion for myocardial bridging, it is recommended that ECG-gated reconstruction be performed during the systolic phase as well as the diastolic phase. Comparison of the images obtained during the two phases will allow assessment of luminal narrowing during the systolic phase. In our study of 956patients, only 3 were found to have the bridging and it was of LAD and of the middle segment with percentage of 0.3.

Duplication of the LAD artery in otherwise normal hearts has been reported to occur in 0.13%–1% of the general population[30]. Duplication of the LAD artery consists of a short LAD artery, which courses and terminates in the anterior interventricular sulcus without reaching the apex, and a long LAD artery, which originates from either the LAD artery proper or the RCA, then enters the distal anterior interventricular sulcus and courses to the apex[30]. Because the LAD artery is the most important coronary artery in coronary artery bypass graft surgery, the cardiologist should be aware of the possibility of duplication of the LAD artery manifesting so that he or she can forewarn the cardiac surgeon of the importance of achieving successful myocardial revascularization; otherwise, there is a risk of incorrect placement of the arteriotomy[31]. Duplication of the LAD artery should not be confused with an LAD artery and a diagonal branch running parallel to each other. Such a parallel diagonal branch does not reenter the anterior interventricular sulcus and take over the course of the distal LAD artery, as does the long anterior LAD artery[32]. We have found one patient having a small LAD terminating only after 2cm and a proper well-formed LAD originating from it and reaching the apex.

Coronary artery ectasia (CAE) or aneurismal coronary artery disease is dilatation of an arterial segment to a diameter at least 1.5 times that of the adjacent normal coronary artery (a). CAE can be found in 3-8% of angiographic and in 0.22% to 1.4% of autopsy series. It can be either diffuse affecting the entire length of a coronary artery, or localized. In Western countries, atherosclerotic aneurysms are most common (50%), followed by congenital (17%) and infectious causes (10%); and in Japan, Kawasaki disease represents the predominant cause of coronary artery aneurysm[14,16,17]. Most of the patients

remain asymptomatic, and aneurysms are incidentally found during diagnostic coronary angiography or at necropsy (q). When patients are symptomatic, the clinical manifestations depend on the underlying cause; in those cases associated with atherosclerosis, the clinical manifestations are similar to those seen in coronary artery disease[21]. The natural history of atherosclerotic aneurysms is usually favorable, but there are conflicting reports[26]. In one study there was no difference in 5-year survival between patients with and without aneurysms who had occlusive coronary artery disease[17]. While in other it was an independent predictor of death, with an overall 5-year survival of 71% and a marked 5-year mortality rate. Nevertheless, no significant associations have been found between aneurysm size and mortality. In our study the ectesia was found to be the most common finding at about the %age of 6.5.

Coronary artery fistula is a condition in which a communication exists between one or two coronary arteries and either a cardiac chamber, the coronary sinus, the superior vena cava, or the pulmonary artery. This condition is seen in approximately 0.1%–0.2% of all patients who undergo selective coronary angiography[33]. It more commonly involves the RCA (60% of cases) than the LCA (40%)[34]. in less than 5% of cases, fistulas originate from both the LCA and the RCA[7].

In coronary artery fistula, the involved coronary artery is dilated because of increased blood flow and is often tortuous to an extent determined by the shunt volume[12]. In terms of morphologic features, the fistula is variable at its drainage site, with either single or multiple communications or a maze of fine vessels that form a diffuse network, or plexus, with extensive intramural distribution. The drainage site of the fistula has a greater clinical and physiologic importance than does the artery of origin. The most common site of drainage is the right ventricle (45% of cases), followed by the right atrium (25%) and the pulmonary artery (15%)[35]. The fistula drains into the left atrium or left ventricle in less than 10% of cases[36]. When the shunt leads into a right-sided cardiac chamber, the hemodynamics resemble those of an extracardiac left-to-right shunt; when the connection is to a left-sided cardiac chamber, the hemodynamics mimics those of aortic insufficiency. Myocardial perfusion may be diminished for that portion of the myocardium supplied by the abnormally connecting coronary artery. This situation represents a hemodynamic steal phenomenon and may lead to myocardial ischemia[37]. We have one young pt of repeated chest pain episodes with otherwise normal ECG and examination.

Coronary artery anomalies may be associated with congenital heart diseases such as transposition of the great arteries, the presence of a single ventricle, dextrocardia, and tetralogy of Fallot[38,39]. The principal purpose of defining coronary artery distribution in congenital heart disease is to forewarn the surgeon of important aberrant epi- or intramyocardial coronary arteries traversing a potential ventriculostomy site.

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