Clinical Investigations

Prevalence of Coronary Artery Anomalies in 12,457 Adult Patients Who Underwent Coronary Angiography

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ABSTRACT

Background: Coronary artery anomalies are found in 0.2% to 1.3% of patients undergoing coronary angiography and 0.3% of an autopsy series. We aimed to estimate the frequency of coronary artery anomalies in our patient population.

Methods: The data were collected retrospectively by analyzing the angiographic data of 12 457 consecutive adult patients undergoing coronary angiography between September 2002 and October 2007.

Results: Coronary artery anomalies were found in 112 patients (0.9% incidence), 100 patients (89.3%) had origin and distribution anomalies, and 12 patients (10.7%) had coronary artery fistulae. Their mean age was 52±8 years (range, 22–79 y). Separate origins of left anterior descending and left circumflex coronary artery from the left sinus of Valsalva was the most common anomaly (63.4%). The right coronary artery rising from the left coronary sinus of Valsalva was found in 10 (8.9%) patients. Anomalous origin of the left circumflex coronary artery from the right sinus of Valsalva was seen in 10 (8.9%) patients. The left main coronary artery from the right coronary sinus of Valsalva was found in 1 (0.89%) patient while an isolated single coronary artery was seen in 2 (1.78%) patients.

Conclusion: The incidence and the pattern of coronary artery anomalies in our patient population were almost identical with previous studies. Cardiologists should be aware of the coronary anomalies which may be associated with potentially serious cardiac events, because recognition of these coronary anomalies is mandatory in order to prescribe appropriate therapy.

Introduction

Congenital coronary artery anomalies are present at birth, but relatively few are symptomatic. Most anomalies are encountered as incidental findings during coronary angiography or at autopsy.[1] About 20% of coronary artery anomalies produce life threatening symptoms including arrhythmias, syncope, myocardial infarction, or sudden death while 80% are benign.[2,3] Coronary artery anomalies are the second most common cause of sudden cardiac death (SCD) in young athletes.[4]

Prevalence of coronary artery anomalies was reported as 0.6% to 1.3% in angiographic series and 0.3% in autopsy series.[1,2,5–7] There are several classifications for coronary artery anomalies. We preferred to use the classification of coronary artery anomalies that was modified to be used in clinical practice in 2 groups; anomalies of origin, distribution, intercoronary communications, and coronary artery fistulae.[5,8,9]

In this trial, our aim was to find the incidence of coronary artery anomalies in patients who underwent coronary angiography for any reason in the Turkish population.

Methods

The data were collected retrospectively by analyzing the angiographic data of 12 457 consecutive adult patients undergoing coronary angiography between September 2002 and October 2007 at Private Gazi Hospital, Izmir, and TDV 29 Mayıs Hospital, Istanbul, Turkey. Coronary angiographies were indicated for stable coronary artery disease in 65% of the patients, acute coronary syndrome in 26% of the patients, valvular heart disease in 7% of the patients, and for other reasons in 2% (congenital heart disease, aortic dissection, etc) of the patients.

The catheterization reports were analyzed, and those with anomalous coronary arteries were selected for further assessment. The angiograms were reviewed by 2 independent investigators before finally being classified. In case of any difference of opinion, a consensus was reached after discussion. Patients with coronary anomalies associated with complex congenital heart disease were not included in this study.

The selective cannulation of aberrant arteries can be difficult and time-consuming. We have used for this

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purpose: hockey-stick, multipurpose, right and left Amplatz, Judkins right and left guide catheters, for angiography and angioplasty.

Results

Coronary artery anomalies were found in 112 patients (incidence of 0.9%). Among the 112 patients (70 males, 42 females), 100 patients (89.3%) had anomalies of origin and distribution and 12 patients (10.7%) had coronary artery fistulae (Table 1). Their mean age was 52±8 years (range, 22–79 y).

The left main coronary artery (LMCA) was the most common anomalous vessel. In this group separate origins of left anterior descending coronary artery (LAD) and left circumflex coronary artery (LCX) from the left coronary sinus of Valsalva (71 patients, 63.4%) was the most common anomaly. Separate origins of LAD and LCX from the right coronary sinus of Valsalva was seen in 1 patient (0.89%).

The right coronary artery arising from the left coronary sinus of Valsalva was found in 10 patients (8.9%). Ectopic origin of the right coronary artery (RCA) from the ascending aorta was seen in 5 patients (4.46%). Anomalous origin of the LCX from the right sinus of Valsalva was seen in 10 (8.9%) patients. The LMCA from the right coronary sinus of Valsalva was found in 1 patient (0.89%) while an isolated single coronary artery was seen in 2 patients (1.78%). The findings of the coronary anomalies were coincidental in our study.

Discussion

Most of the coronary artery anomalies are asymptomatic. The prevalence of coronary artery anomalies shows a wide variation. They are usually encountered as coincidental findings during coronary angiography or at autopsy. In different trials, the incidence was reported as 0.6% to 1.3%. Similarly, we found 0.9% incidence of coronary artery anomalies among patients undergoing diagnostic coronary angiography. The limitation in our study is that only the patients who had undergone coronary angiograms were included rather than a randomly selected sample of the whole population.

Table 1. Incidence of Coronary Artery Anomalies

	No.	Angiographic Incidence (%)	Anomaly Incidence (%)
Total coronary angiograms	12 457	–	–
Total coronary anomalies	112	0.9	–
Anomalies of origin and distribution	100	0.80	89.3
Coronary artery communications and fistulae	12	0.09	10.7

Coronary artery anomalies could lead to life threatening symptoms, including arrhythmias, syncope, myocardial infarction, or sudden death. In 1 report, 33% of patients with anomalous coronary anatomy had a preceding history of angina and or syncope.[2] In our study population, 31% of the patients had typical cardiovascular symptoms.

The origin and distribution of anomalies of the coronary arteries in our study was 89.3% and coronary artery fistulae 10.7%, resembling previous similar studies (95% and 5%, respectively).[2,8,10] The most common anomaly in our study group was separate ostia of LAD and LCX in the absence of LMCA (64.29%) similar to previous trials.[2,10] LCX and RCA anomalies were the second most common anomaly.

An anomalous RCA was the second most common anomaly seen in 15 (13.36%) patients (RCA from the left coronary sinus of Valsalva in 10 patients and RCA from the ascending aorta in 5 patients). In 2 different studies similar to our study, 1 of them from Turkey, the incidence of RCA originating from the left coronary sinus of Valsalva or LAD was reported between 8% to 16%.[10,11]

This was followed by LCX originating from the right coronary sinus of Valsalva in 10 (8.9%) patients. This anomaly was seen in 11% and 18.4% of all coronary anomalies in similar studies from Turkey and central Europe.[8,10]

The anomalous origin of the LMCA from the right coronary sinus of Valsalva is a rare congenital coronary anomaly.[2,5–7] The left main coronary artery from the left coronary sinus of Valsalva was found in 0.017% of patients.[2] We identified 1 (0.008%) patient with this anomaly among 12 457 patients.

The LMCA, LAD, and RCA may arise from the pulmonary artery in the order of decreasing incidence. Unfortunately, about 90% of patients with these anomalies die during infancy.[10] We had no patients with these anomalies because only adult patients were recruited in our trial.

The incidence of the LCX arising from the RCA is generally believed to be of no clinical significance due to a dorsal course the left ventricle.[12]

The LMCA or LAD arising from the right coronary sinus of Valsalva and RCA originating from the left coronary sinus of Valsalva deserve clinical attention because these anomalies may be associated with sudden cardiac death in otherwise fit individuals.[12–16] There were 11 patients with this kind of anomaly in our study population (LMCA from right sinus of valsalva [RSV] in 1 patient, RCA from the left coronary sinus of Valsalva in 10 patients).

The anomalous origin of the LMCA from the right coronary sinus of Valsalva can be subclassified into 4 types based on the relationship of the LMCA to the great vessels: septal (beneath the right ventricular infundibulum), anterior to the pulmonary trunk, retroaortic, and interarterial (between the pulmonary trunk and the aorta). The interarterial course

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Table 2. Distribution of the Coronary Artery Anomalies in 12,457 Adult Patients Who Underwent Angiography

	No.	Incidence (%)	Anomalies (%)
Anomalies of origin and distribution	100	0.80	89.3
Ectopic origin from the sinuses of Valsalva of the aorta	–	–	–
LMCA from RSV	1	0.008	0.89
Separate origins of LAD and LCX in the RSV	1	0.008	0.89
Separate origins of LAD and LCX in LSV	71	0.6	63.4
LCX from RCA or RSV	10	0.08	8.9
RCA from LSV	10	0.08	8.9
Ectopic origin from the ascending aorta	–	–	–
LMCA from ascending aorta	–	–	–
RCA from ascending aorta	5	0.04	4.46
Ectopic origin from the pulmonary artery	–	–	–
Single coronary artery	2	0.016	1.78
RCA supplying the heart	–	–	–
LMCA supplying the heart	–	–	–
Origin of the LAD and LCX from RCA	2	0.016	1.78
Origin of the RCA from the LMCA	–	–	–
Intercoronary communications and coronary artery fistulae	12	0.09	10.7
Intercoronary communications			
AV-node branch-LCX anastomosis	–	–	–
LAD-PDA anastomosis	–	–	–
Coronary artery fistulae	12	0.09	10.7
LAD-PA fistula	7	0.06	6.3
LCX-PA fistula	2	0.02	1.8
Conus branch-RV fistula	1	0.008	0.9
RCA-RV fistula	–	–	–
LCX-RV fistula	1	0.008	0.9
RCA-RA fistula	1	0.008	0.9

Abbreviations: AV = atrioventricular; LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery; LMA = left main coronary artery; PA = pulmonary artery; PDA = posterior descending coronary artery; RA = right atrium; RCA = right coronary artery; RSV = right sinus of valsalva; RV = right ventricle.

was found in 60% of patients in whom the left main coronary artery originates from the right coronary sinus.[17] Patients with this form of LMCA anomaly have complaints of chest discomfort and suffer from premature cardiac death.[4,15]

Reduction of the coronary blood flow resulting from compression over LMCA from the right coronary sinus of Valsalva or the first segment of a single coronary artery such as the RCA from the left coronary sinus of Valsalva or LAD,

coursing interarterially, by the pulmonary trunk or aorta during pressure increase in these vessels upon exercise in addition to acute takeoff or slit-like orifices of these arteries, produce ischemia resulting in angina, syncope, congestive heart failure, arrhythmias, and sudden death.[15,18–22]

Until recently, the exact anatomic location has been shown during catheterization by means of the "dot and eye" method and/or placement of a catheter in the pulmonary artery.[17]

Computed tomographic angiography is a safe and effective noninvasive imaging modality for defining coronary anomalies and provides detailed 3-dimensional anatomic information that may be difficult to obtain with invasive angiography.[12]

The treatment of these coronary anomalies is controversial. Very few cases of percutaneous transluminal coronary angioplasty in anomalous arteries have been reported.[23–27] Basically the 3 benign forms of the anomalously originating LMCA from the right coronary sinus of Valsalva do not require specific interventions or cessation of sports activities, while interarterially coursing LMCA may warrant surgical repositioning, especially if associated with objective evidence of ischemia.[22]

It is important to differentiate this interarterial course from intraseptal (which may appear similar, but the anomalous vessel passes more inferiorly within the muscular septum) and the posterior course that have relatively benign courses than the first one.[12] Because of the unusual location and the noncircular coronary orifice of these anomalies, selective catheterization and percutaneous intervention can be technically challenging, particularly with regard to adequate guide catheter support.

Coronary artery fistulas (CAF) are rare congenital anomalies.[28] The incidence is around 0.002% in the general population and 0.3% to 0.4% in patients with congenital heart defects.[29,30] The CAF incidence in our study was 0.09% among patients undergoing diagnostic coronary angiography. Coronary artery fistulas consist of a communication between a coronary artery and a cardiac chamber or a great vessel. In CAF, blood diverting from the high resistance myocardial capillary bed into the low resistance fistula produce ischemia and coronary steal phenomenon.[31] The most common type of fistula is the coronary to pulmonary artery type and usually arises from the LAD or the RCA. Early phase CAF is generally not symptomatic, but later on it can cause important coronary morbidity and mortality leading to angina, syncope, congestive heart failure, myocardial infarction, and sudden death. Surgical ligation is the standard treatment for symptomatic CAF, but is associated with higher morbidity.[32] In the last decade, percutaneous methods have developed as an alternative treatment modality. We treated 2 cases in whom myocardial ischemia was due to coronary steal by using covered stent and coil occlusion.

In conclusion, coronary artery anomalies are rarely identified during life, often because of insufficient clinical suspicion. Familiarity with coronary artery anomalies may be useful for physicians dealing with diagnosis and treatment of these pathologies.

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