Coronary Artery Abnormalities in Puerto Rico

YOLANDA FIGUEROA, MD; PABLO I. ALTIERI, MD; HÉCTOR BANCHS, MD; NELSON ESCOBALES, Ph D; MARÍA CRESPO, PhD; EFRAÍN DEFENDINI, MD; IVÁN GONZÁLEZ-CANCEL, MD

A retrospective study was done to determine the frequency of coronary artery anomalies in terms of their origin, course, and structure. The clinical history, catheterization data and surgical reports of patients undergoing coronary angiography at the Cardiovascular Center of Puerto Rico and the Caribbean, from 1999 to 2004, were analyzed. Thirty-eight patients were identified with a coronary artery anomaly in this population. These anomalies were classified according to their clinical consequences and the need for surgical intervention.

Key words: Coronary artery abnormalities, Coronary circulation, Ectopic coronary arteries, Coronary artery malformations, Angiography

The coronary arteries may present a wide range of variations in terms of origin, course, and structure. Usually these variations are discovered incidentally during coronary angiography or at autopsy. According to literature the incidence of all coronary anomalies ranges between 0.3% and 12% in angiographic series (Chaitman et al., 1976 [1]; Engel et al., 1975 [2]; Barriales et al., 2001[3]) and 0.23% in autopsy series (Alexander and Griffith, 1956[4]). Most coronary artery anomalies do not affect the quality of life or lifespan of the affected individuals, but some have been implicated in the development of chest pain, cardiomyopathy, dyspnea, syncope, ventricular fibrillation, myocardial infarction, and sudden death. By determining the incidence of anatomic variants in a large population, acceptable definitions of normal and anomalous anatomy have been established and the clinical relevance of anomalous variants ascertained. "Benign anomalies" are identified as 1) separate origin of the left anterior descending and circumflex from the left sinus of Valsalva; 2) ectopic origin of the circumflex from the right sinus of Valsalva; 3) ectopic coronary origin from the posterior sinus of Valsalva; 4) anomalous coronary origin from the ascending aorta; 5) absent circumflex; 6) intercoronary communications; and 7) small coronary artery fistulae. The coronary artery anomalies that have been associated with potentially serious sequelae include: 1) ectopic coronary origin from the pulmonary artery; 2) ectopic coronary origin from the opposite aortic sinus; 3) single coronary artery; and 4) large coronary fistulae. Coronary artery anomalies require accurate recognition for proper evaluation, management, and at times, surgical correction [5-9].

Departments of Medicine, Physiology, and Surgery, University of Puerto Rico, Medical Science Campus, Cardiovascular Center of Puerto Rico and the Caribbean, Ramón M. Suárez Calderón

Address correspondence to: Pablo I. Altieri, MD, Box 8387, Humacao, PR 00792

Materials and Methods

The database of the cardiac catheterization laboratories of Cardiovascular Center of Puerto Rico and the Caribbean was used in this retrospective review. The clinical history, catheterization data and surgical reports of the patients undergoing coronary angiography from 1999 to 2004 were obtained. This data was analyzed in order to identify the anomalous origins, courses, and communications between coronary arteries in this population, and to classify them in terms of their consequences and the need for surgical intervention.

Results

Among the patients who underwent coronary angiography between 1999 and 2004 at the CCPRC, the mean age of patients identified with coronary artery anomalies was 44 years with a range of 1 year to 87 years of age. Forty seven percent were males and 53% females (Table 1). A total of 38 patients were identified with a coronary artery anomaly in terms of its origin, course, or structure (Table 2). In 4 cases (11%) the right coronary

Table 1. Study Population

Age	Sex	Symptoms	EF %	Anomaly	Other Lesions	Intervention
55	F	Angina	55	Lt main - PA fistula	Non obstructive disease	Suture ligation
23	F	STEMI	60	RCA - septum fistula	Free of disease	Medical Tx
56	F	Angina	30	LAD - PA fistula	4 vessels (80-100%)	PTCA
65	M	Angina	60	LAD - PA fistula	Non obstructive disease	Suture ligation
4	M	Tachy	68	LAD - LV fistula	Free of disease	Suture ligation
7	F	Asympt	66	RCA - RV fistula	Free of disease	Suture ligation
1	F	Cyanosis	44	LCA - LV fistula	PDA	Ligation + PDA closure
57	F	Asympt	55	LCA - PA fistula	60% LAD	Suture ligation
53	M	Angina	55	RCA from Lt sinus	Free of disease	Medical Tx
56	F	Angina	70	RCA from Lt sinus	Free of disease	RCA ostium translocation into Rt coronary sinus
36	M	Angina	55	RCA from Lt sinus	50-70% RCA prox	RCA ostium translocation into Rt coronary sinus
44	M	Angina	60	RCA from Lt sinus	Free of disease	RCA ostium translocation into Rt coronary sinus
3	F	VSD	68	LAD from RCA	Free of disease	Implantation RV to VA conduit
1	F	CHF	35	LCA from PA	Free of disease	Implantation LCA to Aorta
51	M	Angina	47	CX from Rt cor ost	1° Pulm HTN	Medical Tx
87	F	Asympt	80	Anom origin Lt + CX	Free of disease	Medical Tx
3	M	Asympt	/	Corons from Aorta	VSD	VSD repair
61	F	Angina	60	Hypoplastic RCA	Non obstructive disease	Medical Tx
47	F	Angina	50	Hypoplastic RCA	Free of disease	Medical Tx
67	F	Angina	55	Hypoplastic RCA	2 vessels (50-90%)	CABG
45	M	Asympt	60	RCA prox nicking	Free of disease	Medical Tx
68	F	Angina	70	Abnorm distal LAD	Free of disease	Medical Tx
54	F	Angina	/	LAD anomaly	////////	CABG x1
42	F	Syncope	60	Musl bridge mid LAD	Free of disease	Medical Tx
31	M	NSTEMI	70	Musl bridge mid LAD	Free of disease	Medical Tx
73	F	Angina	70	Musl bridge mid LAD	20% OM1 + OM2	Medical Tx
75	F	Asympt	75	Musl bridge dist LAD	Free of disease	Medical Tx
32	M	ASD	60	Muscle bridge LAD	Free of disease	ASD repair
37	M	Asympt	60	Muscle bridge LAD	Non obstructive disease	Medical Tx
61	M	Angina	60	Muscle bridge LAD	2 vessels (30-50%)	Medical Tx
58	F	NSTEMI	60	Muscle bridge LAD	Non obstructive disease	Medical Tx
64	M	Angina	55	Muscle bridge LAD	2 vessels (30-80%)	Medical Tx
47	M	Angina	40	Muscle bridge LAD	2 vessels (80%) + non obstr	PTCA
48	M	Asympt	70	Muscle bridge LCA	Free of disease	Medical Tx
47	M	Asympt	70	Muscle bridge LCA	40% LCX	Medical Tx
53	M	Angina	50	Musl ectopy CX branch	Non obstructive disease	PTCA + stent
67	M	Old MI	50	Bridging collat RCA	3 vessels (90-99%)	CABG x3
2	F	Asympt	/	Lg aorto-pulm collat	Several collaterals	Coil embolization

*PA, pulmonary artery; RCA, right coronary artery; LCA, left coronary artery; LCX, left circumflex; LAD, left anterior descending; LV, left ventricle; RV, right ventricle; MI, myocardial infarction; ASD, atrial septal defect; VSD, ventricular septal defect; PDA, patent ductus arteriosus; CHF, congestive heart failure.

artery had an anomalous origin from the left coronary sinus. Of these, 3 patients required surgical intervention with translocation of the RCA ostium into the right coronary sinus due to development of chest pain, palpitations, and increasing shortness of breath. An anomalous origin of the circumflex was observed in 2 cases (5.26%), one of them from the right coronary ostium, but none requiring surgery. The left anterior descending artery originated from the right coronary artery in one case (2.6%). Origin of the left coronary artery from the pulmonary artery was identified in one case (2.6%). This patient underwent reimplantation of the LCA to the ascending aorta after presenting with congestive heart failure.

Fistulous communications between a coronary artery and a cardiac cavity or any part of the pulmonary or systemic circulation were identified in 8 cases (21%). Seven of these fistulae (88%) were large enough to require surgical closure. Muscle bridges were observed in the majority of cases (32%), but none of these cases presented with signs, symptoms, or complications attributable to this anomaly, for which they were considered benign.

Discussion

Although coronary artery anomalies are very rare, some of them present with symptoms or potentially serious sequelae that require surgical treatment. In our database, 32% of the patients were found with a coronary artery anomaly that has been associated with a potentially serious outcome. These patients required surgical intervention.

Table 2. Distribution of Coronary Artery Anomalies

Coronary Artery Anomaly	Number	Percentage
Anomalous RCA origin from Left coronary sinus	4	10.50%
Anomalous LAD origin from RCA	1	2.63%
Anomalous LCA origin from PA	1	2.63%
Anomalous CX origin from Right coronary ostium	1	2.63%
Anomalous origin of Left main and CX	1	2.63%
Anomalous Coronaries origin from Ascending Aorta	1	2.63%
Left main to PA fistula	2	5.26%
Left main to LV fistula	1	2.63%
LAD to PA fistula	2	5.26%
LAD to LV fistula	1	2.63%
RCA to Septum fistula	1	2.63%
RCA to RV fistula	1	2.63%
Hypoplastic RCA	3	7.89%
RCA proximal nicking	1	2.63%
Abnormal distal LAD course	1	2.63%
LAD anomaly	1	2.63%
Muscle bridge to LAD	10	26.30%
Muscle bridge to LCA	2	5.26%
Muscle ectopy CX branches	1	2.63%
Bridging collaterals to RCA	1	2.63%
Large Aorto-Pulmonary collateral	1	2.63%

*LAD, left anterior descending; LCA, left coronary artery; RCA, right coronary artery; CX, circumflex; LV, left ventricle; PA, pulmonary artery.

Fifty eight percent of the cases with coronary artery fistulae required suture ligation, mostly due to symptoms of angina. Seventy five percent of those with anomalous RCA origin from the left coronary sinus also presented with symptoms of angina and required translocation of the RCA ostium into the right coronary sinus. Patients affected by this anomaly most frequently present with ischemic complications especially in the presence of atherosclerosis [1,5].

Only one case was found with the origin of the left coronary artery from the pulmonary artery, a one year old patient. This is known as the Bland-White-Garland syndrome in which blood flow originates in a dilated RCA and passes via collaterals to the left coronary artery where it flows in a retrograde fashion into the pulmonary artery. Ninety percent of patients die in infancy and very few survive to reach adulthood, according to literature [1,5]. The high risk of mortality in these patients warrants the immediate correction of the anomaly when diagnosed.

Sudden death is frequently the only symptom of an anomaly. Understanding of the variability in anatomy, pathophysiology, and functional repercussions of such anomalies is lacking. This should be a healthcare priority in regard of proper screening, clinical recommendations, prevention, and treatment. The clinician should suspect the presence of coronary artery anomalies especially in the young patient presenting with exertional syncope, myocardial infarction, exercise induced arrhythmias, or cardiac arrest. Proper identification and classifications of these anomalies is essential for adequate evaluation and avoid errors in medical or surgical management. Long term observation and follow up of these patients should be considered for evaluation of the anomalies' natural history and the outcomes of any intervention [1-5].

Resumen

Se realizó un estudio retrospectivo para determinar la frecuencia de anomalías en términos de origen, trayectoria y estructura de las arterias coronarias. Los expedientes de pacientes con angiografías coronarias realizadas en el Centro Cardiovascular de Puerto Rico y el Caribe en el periodo desde el 1999 al 2004 fueron evaluados. Se analizaron los historiales clínicos, reportes de cateterismo y reportes quirúrgicos en esta población. Un total de 38 pacientes fueron identificados con anomalías en las arterias coronarias. Estas anomalías fueron clasificadas de acuerdo a sus consecuencias clínicas y la necesidad de intervención quirúrgica en estos pacientes.

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