

Anomalous Coronary Arteries Originating in the Contralateral Sinus of Valsalva: Registry of Thirteen Spanish Hospitals (RACES)

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The registry contains information collected during 2003 from 13 Spanish hospitals on the incidence, clinical presentation, initial course, diagnostic imaging, and treatment of anomalous coronary arteries originating in the contralateral coronary sinus. In a total of 23 300 coronary angiograms, 98 anomalous coronary arteries were detected (incidence 0.4%): 46 circumflex coronary arteries (47%), 25 right coronary arteries (25.5%), 10 left main coronary arteries (10.2%), 3 left anterior descending arteries (3.1%), 6 single coronary arteries (6.1%), and 8 other anomalies (8.2%). The most frequent indication for angiography was angina (43.9%). The initial course of the anomalous coronary artery was not identified in 40.8% of cases. In 2 patients (2%), diagnostic imaging was used to identify the initial course. Associated atherosclerotic disease was present in 51% of patients. In addition, 18.4% had associated valvular disease, with the aortic valve being affected most often (15.2%). Some 16.3% of patients underwent treatment for the coronary anomaly. The incidence was similar to that observed in other published series, with anomalies in the circumflex coronary artery being diagnosed most frequently. Diagnostic imaging was underused.

Key words: Angiography. Coronary angiography. Congenital heart defects. Congenital defects. Imaging.

Registro de anomalías congénitas de las arterias coronarias con origen en el seno de Valsalva contralateral en 13 hospitales españoles (RACES)

Este registro estudió la incidencia, la clínica asociada, el trayecto, los métodos diagnósticos de imagen y el tratamiento de las anomalías coronarias con origen en el seno coronario contralateral en 13 hospitales durante 2003. En 23.300 coronariografías se describieron 98 anomalías (incidencia del 0,4%): 46 circunflejas (47%), 25 coronarias derechas (25,5%), 10 coronarias izquierdas (10,2%), 3 descendentes anteriores (3,1%), 6 coronarias únicas (6,1%) y 8 incluidas en el apartado «otras» (8,2%). El motivo de la coronariografía más frecuente fue la angina (43,9%). El trayecto inicial de la coronaria anómala no fue identificado en el 40,8% de los casos. En 2 pacientes (2%) se emplearon métodos de imagen asociados. El 51% tenía enfermedad coronaria asociada y el 18,4%, valvular; la válvula aórtica era la más afectada (15,2%). El 16,3% recibió tratamiento para la anomalía. Se observó una incidencia similar a la de series previas, y las anomalías de la circunfleja fueron las más frecuentes. Los métodos de imagen asociados para el diagnóstico del trayecto inicial fueron infrutilizados.

Palabras clave: Angiografía. Coronariografía. Defectos cardíacos congénitos. Defectos congénitos. Imagen.

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INTRODUCTION

The considerations regarding congenital anomalies of the coronary arteries has shifted in recent years. From simple coronary angiography findings of minor clinical importance, they are now known to be the cause of sudden death in youths and a possible cause of ischemia in the adult population.^{1,2}

International forums have been calling for the creation of registries that include the group of coronary anomalies (CAs), to which are attributed most of the

clinical events (sudden death and myocardial ischemia) occurring among the young and adult populations.³ Specifically, these anomalies include those originating in the contralateral sinus of Valsalva.

In order to describe this type of CA in a group of Spanish hospitals, the Registry of Coronary Artery Congenital Anomalies Originating in the Contralateral Sinus of Valsalva (RACES, *Registro de Anomalías Congénitas de las Arterias Coronarias con Origen en el Seno de Valsalva Contralateral*) was created among a group of 13 Spanish hospitals.

METHODS

The data recorded included congenital abnormalities originating in the contralateral sinus of Valsalva as well as single coronary arteries because these sometimes establish courses similar to those of CAs originating in the contralateral sinuses.

This was a prospective study in which each investigator, prior to the coronary angiography diagnosis of coronary anomaly, completed a questionnaire that included: patient age and sex, symptom for which the study was performed, description of the CA and its initial course, other diagnostic imaging methods used, presence of associated coronary disease (considered significant if >50% of total vessel lumen) and/or valvular disease, and treatment. Each hospital followed its usual diagnostic protocols. Informed consent was requested from each patient to be included. The study was conducted from 1 January to 31 December 2003.

RESULTS

Among 23 300 coronary angiographies, 98 CAs (incidence, 0.4%) were found to originate in the contralateral sinus (Table 1). Among the total studied,

70% were men (16 317) and 30%, women (6983). Among patients with CA, 70 were men (71%) and 28 women (29%). The mean age was 61.03 ± 14.39 years (range, 16-87 years).

The most frequent indication for coronary angiography was angina (43.9%), followed by valve disease (20%), atypical pain (17.3%), acute myocardial infarction (14.3%), dyspnea (11.2%), syncope (1%), and dizziness (1%). No relationship was found between the type of CA and the presentation.

Forty-six circumflex arteries (Cx) (47%) originated in the right sinus of Valsalva (RSV) or right coronary artery (RCA), 25 RCA (25.5%) arose from the left sinus of Valsalva (LSV) (Figure 1), 10 left coronary arteries (LCA) (10.2%) originated in the LSV, 3 anterior descending arteries (3.1%) in the RSV, and 6 single coronaries (6.1%) and 8 CAs (8.2%) were included in the group of *others* (the anomaly was anatomically complex and could not be included in the above groups, but was characterized as originating in the contralateral sinus).

The initial course of the anomalous coronary was retroaortic in 36 (36.7%), anterior in 12 (12.2%), interarterial in 10 (10.2%), and "unknown" (or unidentified) in 40 patients (40.8%). The arterial course was diagnosed according to coronary angiography criteria (98%), with additional imaging used as a diagnostic method in only 2 cases.

Among the total of CA patients, 50 patients (51%) had associated coronary disease (in any coronary artery), but there was no relationship between the type of anomaly and the presence of coronary disease. Associated valve disease was found in 18%, and a statistically significant relationship was found between the presence of CA and aortic valve disease.

The CA was treated in 16.3% patients, most of them (15 patients) because of the presence of associated coronary disease. These cases were treated as any

TABLE 1. Hospitals and Investigators Participating in the RACES Registry. Total Number of Coronary Angiographies During 2003 and Number of Anomalies Described*

Hospital	Investigator	Coronary Angiographies,n	CA,n
Hospital Meixoeiro (Vigo)	Marcelo Sanmartín	2888	6
Hospital General Universitario de Alicante (Alicante)	Pascual Bordes	2663	8
Complejo Universitario Juan Canalejo (La Coruña)	Jorge Salgado	2456	6
Hospital Universitario Central de Asturias (Oviedo)	César Morís	2434	17
Policlínica Guipúzcoa (San Sebastián)	Juan C. Sanmartín	2273	11
Hospital de Bellvitge (L'Hospitalet de Llogregat)	Emili Iráculis	2090	8
Hospital Universitario Virgen de la Salud (Toledo)	Fernando Pajín	1996	9
Hospital Universitario Marqués de Valdecilla (Santander)	J.M. de la Torre	1650	8
Hospital de León (León)	Felipe Fernández	1223	2
Hospital San Juan de Alicante (Alicante)	J.M. Ruiz Nodar	1105	9
Hospital German Trias i Pujol (Badalona)	Eduard Fernández	1083	10
Hospital de Galdakao (Galdakao))	J.R. Rumoroso	789	3
Hospital Universitario Dr. Peset (Valencia)	Adolfo Rincón	640	1

*CA indicates coronary anomalies.

other atherosclerotic lesion, with no special considerations related to their anomalous course. Only one patient underwent surgery because of the anomaly.

DISCUSSION

The incidence in our registry was 0.4%, which is similar to previously published data.^{2,4-7} It is worth noting the difference in the number of CAs described between hospitals with a similar number of coronary angiographies (Table 1). We do not believe this is the result of any inherent differences between the populations in the autonomous communities of Spain. As has been observed by Angelini et al, the number of CAs described in each laboratory varies considerably due to the “meticulousness” of each group in the diagnosis.^{1,3}

Once corrected for the total number of men and women undergoing catheterization, we found no gender-related differences in the incidence of CAs. As in most published studies, the Cx anomalies in our series (arising from the RSV or the origin of the RCA) were the most frequent^{2,8} (47%) (Figure 2).

The initial course of the anomalous coronary, i.e., its anatomical relationship with the aorta and pulmonary artery, is an essential aspect when describing a CA, because certain courses (e.g., interarterial) are associated with a higher incidence of ischemia.^{9,10} One of the most interesting aspects of our registry was the almost exclusive use of coronary angiography to describe the initial courses. This may be the reason why the initial course was not identified in most patients (40.8%). Coronary angiography gives us a two-dimensional view of the coronary vasculature, a complex three-dimensional structure; hence other imaging methods are used to confirm or diagnose these courses (e.g., transthoracic or transesophageal echocardiography, magnetic resonance, or computed tomography).¹¹⁻¹⁴ Nevertheless, in our registry, an additional technique was used in only 2 patients: transesophageal echocardiogram in one and magnetic resonance in the other. The course identified most often was retroaortic (36.7%), because of the predominance of anomalous circumflex arteries (these arteries tend to follow this course).²

The possible predisposition of the anomalous course of the CA to develop atherosclerotic disease was and is one of the more controversial points in CA studies. In fact, there appears to be an equal number defending each of the theories.^{2,8,15,16} In our registry, we observed associated coronary disease (in any coronary) in 51% of the cases, a finding consistent with the fact that most patients were referred for a study of ischemic heart disease with angina as the predominant symptom. However, we found no relationship between the presence of atherosclerotic disease and the different types of CAS (Table 2).



Figure 1. Coronary angiography showing a right coronary artery originating in the left sinus of Valsalva.

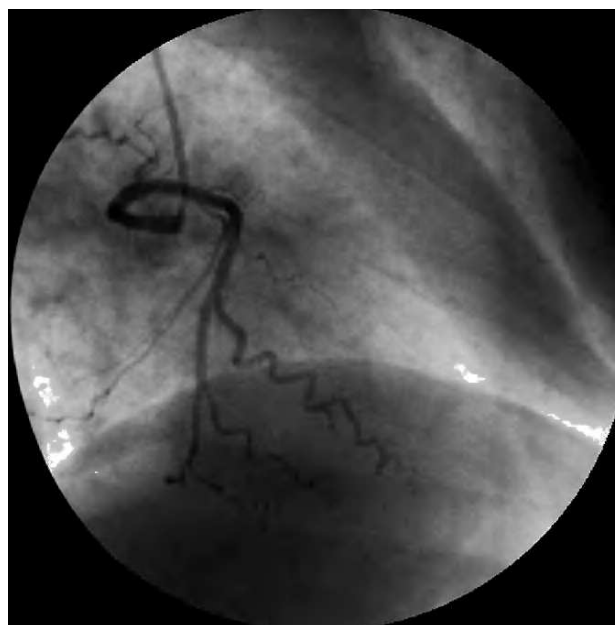


Figure 2. Coronary angiography showing a circumflex artery that originates in the right sinus of Valsalva and takes a retroaortic course until reaching its point of normal distribution.

Aortic valve disease (primarily bicuspid) is known to be associated with CA, and this fact should be taken into account when considering potential surgery for this type of patient.¹⁷

Limitations of the Study

A specific joint protocol for CA diagnosis was not used in this registry. As a result, coronary angiograms were not analyzed by a core laboratory, but rather, different criteria were applied when establishing the diagnosis and describing the anomalous courses.

TABLE 2. Relationship Between Type of Coronary Anomaly and Location of Associated Coronary Lesions*

Associated Coronary Lesions		Type of Coronary Anomaly						Total
		RCA	LCA	Cx	LAD	Single	Other	
Arterial territory affected	None	10	7	20	2	4	5	48
	LAD	3	0	5	0	1	1	10
	LAD, Cx	0	0	4	0	0	0	4
	LAD, Cx, RCA	2	0	2	0	0	1	5
	LAD, Cx, RCA, LMCA	0	0	1	0	0	0	1
	LAD, RCA	4	0	1	1	0	0	6
	Cx	2	1	5	0	1	0	9
	RCA, Cx	0	0	5	0	0	0	5
Total	RCA	4	2	3	0	0	1	10
		25	10	46	3	6	8	98

*Cx indicates circumflex artery; LAD, left anterior descending artery; LCA, left coronary artery; LMCA, left main coronary artery; RCA, right coronary artery.

Lastly, the use of additional imaging techniques was left to the discretion of each investigator.

The incidence of CA is similar to that reported in other series, showing a between-hospital variation possibly due to differences in “meticulousness” in the diagnosis of each group. There were no differences between men and women, and Cx anomalies were the most common. The initial course of the CAs was not identified in most cases, and an additional imaging technique was used in only 2 patients to diagnose this course. One patient was treated surgically only for the anomaly. We did not find a higher incidence of atherosclerosis in the anomalous courses, but did find an association between aortic valve disease and the presence of CA.

The RACES results are likely to indicate that we are not sufficiently aware of the potential prognostic importance of this condition because of its low incidence. We believe these data represent an extremely important starting point for the future establishment of diagnostic standards and protocols for CAs.

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