

Anomalous origin of the right coronary artery from the left sinus of Valsalva: prevalence, clinical characteristics, and long-term follow-up

Origem anômala da artéria coronária direita no seio de Valsalva esquerdo: prevalência, características clínicas e seguimento de longo prazo

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ABSTRACT – Background: The anomalous origin of coronary arteries is uncommon in patients undergoing coronary angiography. In some cases, in their course, they may be subjected to extrinsic compression, which results in myocardial ischemia. The present study aimed to report the prevalence, clinical features and prognostic impact of the anomalous origin of the right coronary artery from the left sinus of Valsalva. **Methods:** We tracked down the medical records of the procedures performed in an interventional cardiology service, between 1999 and 2012. By reviewing medical records and by telephone calls, we obtained the clinical characteristics and carried out a follow-up of the patients with anomalous origin of the right coronary artery from the left sinus of Valsalva. **Results:** Out of a total of 133,277 procedures, the prevalence of this anomaly was 0.09% (n=129) - 36 of these (27.9%) had an interarterial course. The mean age was 58.3±12.4 years. The clinical presentation ranged from asymptomatic patients to acute coronary syndrome, and only two individuals (1.6%), aged 29 and 46 years, were admitted with reversed sudden death. There were 29 (64.4%) and 4 (5.3%) percutaneous or surgical revascularizations, respectively, with and without significant atherosclerotic involvement of the right coronary artery. After a mean follow-up of 1,620 days, the cardiac mortality was 4.6%. **Conclusions:** An anomalous origin of the right coronary artery from the left sinus of Valsalva is an uncommon finding in patients undergoing coronary angiography. Revascularization is often due to a concomitant atherosclerotic involvement of the vessel, and the mortality rate in late follow-up is low and similar to that found in the general population.

Keywords: Cardiovascular abnormalities; Coronary circulation; Cardiac catheterization

RESUMO – Introdução: Anomalias de origem das artérias coronárias são achados pouco frequentes em pacientes submetidos à cinecoronariografia. Em alguns casos, em seu trajeto, elas podem sofrer compressão extrínseca, ocorrendo isquemia miocárdica. O presente estudo teve como objetivo relatar a prevalência, as características clínicas e o impacto prognóstico da origem anômala da artéria coronária direita oriunda do seio de Valsalva esquerdo. **Métodos:** Foi feito um rastreamento nos procedimentos realizados em um serviço de cardiologia intervencionista, no período entre 1999 e 2012. Por meio de revisão dos prontuários e contato telefônico, foram obtidas as características clínicas, tendo sido realizado o seguimento dos indivíduos com origem anômala da artéria coronária direita oriunda do seio de Valsalva esquerdo. **Resultados:** Em um total de 133.277 procedimentos, a prevalência desta anomalia foi de 0,09% (n=129) - 36 destas (27,9%) com trajeto interarterial. A idade média foi de 58,3±12,4 anos. A apresentação clínica abrangeu um espectro que variou desde os assintomáticos até a síndrome coronariana aguda, e somente dois (1,6%) pacientes, de 29 e 46 anos, foram admitidos com morte súbita revertida. Ocorreram 29 (64,4%) e 4 (5,3%) revascularizações percutâneas ou cirúrgicas, respectivamente, com ou sem acometimento aterosclerótico significativo da artéria coronária direita. Após 1.620 dias de seguimento médio, a mortalidade cardíaca foi de 4,6%. **Conclusões:** A origem anômala de artéria coronária direita no seio de Valsalva esquerdo é achado pouco frequente em pacientes submetidos à cinecoronariografia. A revascularização é, em geral, motivada por acometimento aterosclerótico concomitante do vaso, e a mortalidade no seguimento tardio é baixa e próxima à da população geral.

Descritores: Anormalidades cardiovasculares; Circulação coronária; Cateterismo cardíaco

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INTRODUCTION

Anomalous origin of the coronary arteries are found in approximately 0.6% to 1.3% of patients undergoing coronary angiography.¹ Most patients remain asymptomatic throughout life, but some develop symptoms related to myocardial ischemia. The clinical relevance of these anomalies, however, is due to the possibility that sudden death may be their first manifestation, and an anomalous origin of the coronary artery is the second most common cause of sudden death in young athletes.^{2,3}

Cardiovascular events are most frequently related to coronary arteries originating from the contralateral sinus of Valsalva, especially those with a course between the aorta and the pulmonary artery (interarterial). In this group, we found an estimated 0.019% to 0.92% incidence of anomalous origin of the right coronary artery (RCA) from the left sinus of Valsalva, among the patients who underwent coronary angiography,^{4,5} with serious prognostic implications in young individuals.^{6,7}

The objective of this study was to report the incidence of this anomaly, and to describe the clinically relevant characteristics of these patients and their long-term follow-up.

METHODS

We tracked down the medical records in the database of the Catheterization Laboratory and Interventional Cardiology Department of *Instituto do Coração of Hospital das Clínicas de Faculdade de Medicina of Universidade de São Paulo (USP)*, in the city of São Paulo (SP, Brazil), from 1999 to 2012, using the terms “anomalous origin”, “anomaly of origin” and “interarterial course”. The study was submitted to the Institutional Review Board and it approved (CAEE: 92024418.4.0000.0068).

After ruling out other non-relevant diagnoses, we found 239 patients with anomaly of origin of the RCA, 110 of whom were excluded due to origins other than the left sinus of Valsalva, or because we could not obtain reliable data on the medical record.

We obtained the clinical characteristics and follow-up information of the patients by reviewing the medical records and by telephone calls. There was no loss of follow-up regarding any of the 129 patients included in the study.

Statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS), version 22.0 (SPSS Inc, Chicago, USA). The categorical variables were expressed in absolute frequency and percentages, and the continuous variables, as mean and standard deviation.

RESULTS

Between 1999 and 2012, 133,277 coronary angiographies were performed in the service, of which 239 had a diagnosis of anomalous origin of the RCA (prevalence of 0.17%), 129 with confirmed origin from the left sinus of

Valsalva (prevalence of 0.09%); 36 of whom (27.9%) had an interarterial course.

The mean age of patients was 58.3±12.4 years; 57.4% were male, and most of them had risk factors for coronary artery disease (Table 1).

The clinical presentations ranged from asymptomatic patients to acute coronary syndrome, including other symptoms such as dyspnea, heart palpitations, or syncope. Only two (1.6%) patients, aged 29 and 46 years, were diagnosed as reversed sudden death.

Table 1. Clinical, anatomical and functional characteristics

Variable	n=129
Age, years	58.3±12.4
Male	74 (57.4)
Diabetes	40 (31.0)
Hypertension	97 (75.2)
Family history of early CAD	3 (2.3)
Dyslipidemia	63 (48.8)
Previous myocardial infarction	8 (6.2)
Clinical presentation	
Asymptomatic	14 (10.9)
Stable angina	46 (35.7)
NSTEMI not related to RCA	24 (18.6)
NSTEMI related to RCA	4 (3.1)
STEMI not related to RCA	4 (3.1)
STEMI related to RCA	3 (2.3)
Syncope	3 (2.3)
Dyspnea	22 (17.1)
Reversed sudden death	2 (1.6)
Palpitations	7 (5.3)
Functional assessment of ischemia	
No assessment	71 (55.0)
No ischemia	44 (34.1)
Anterior wall ischemia	4 (3.1)
Inferior wall ischemia	8 (6.2)
Lateral wall ischemia	2 (1.6)
Arterial pattern	
No CAD	47 (36.4)
One-vessel	33 (25.6)
Two-vessel	22 (17.1)
Three-vessel	27 (20.9)
Involvement of the RCA	
No CAD/mild CAD	75 (58.1)
Moderate CAD	7 (5.4)
Significant CAD	42 (32.6)
Moderate compression	2 (1.6)
Significant compression	3 (2.3)
Interarterial course	36 (27.9)
Ejection fraction	60.0

Results expressed as mean ± standard deviation, n (%) and %. CAD: coronary atherosclerotic disease; NSTEMI: non-ST segment elevation myocardial infarction; RCA: right coronary artery; STEMI: ST segment elevation myocardial infarction.

Functional tests were performed on 58 patients, and only 8 (6.2%) showed inferior wall ischemia. Of these, 4 cases were related to atherosclerotic vessel involvement, and 4 patients had normal coronary arteries.

Of the total, 45 patients (34.9%) had significant RCA involvement, 42 of whom had coronary artery disease and 3 had significant extrinsic compression, 16 (35.5%) remained in clinical treatment, 13 (29%) underwent surgical revascularization, and 16 (35.5%) underwent angioplasty with stenting (Table 2).

Out of 75 patients (58.1%) with no RCA involvement, 4 (5.3%) underwent coronary artery bypass graft (CABG)

- 2 due to stable angina as presenting symptom with scintigraphic evidence of inferior wall ischemia, 1 for reversed sudden death (29-year-old patient), and 1 patient who was revascularized with no objective evidence of ischemia, based on typical angina symptoms.

The mean follow-up after diagnosis was 1,620 days, and the all-cause mortality rate in this group was 9.3%. Of the 12 deaths, 6 were from non-cardiac causes, and 6 (4.6%) were from cardiac causes (2 patients without previously detected coronary disease, who were in clinical treatment, and 4 patients who had significant coronary disease and also underwent clinical treatment, 2 of whom due to angioplasty failure).

Table 2. Involvement of the right coronary artery (RCA) and respective treatment

RCA treatment	CAD in the RCA					Total
	No CAD	Moderate CAD	Moderate compression	Significant CAD	Significant compression	
No treatment	71 (94.7)	7 (100)	2 (100)	14 (33.3)	2 (66.7)	96 (74.4)
Successful PCI	0	0	0	11 (26.2)	0	11 (8.5)
Unsuccessful PCI	0	0	0	5 (11.9)	0	5 (3.9)
CABG	4 (5.3)	0	0	12 (28.6)	1 (33.3)	17 (13.2)
Total	75 (100)	7 (100)	2 (100)	42 (100)	3 (100)	129 (100)

Results expressed as n (%). CAD: coronary atherosclerotic disease; PCI: percutaneous coronary intervention; CABG: coronary artery bypass graft

DISCUSSION

As far as we know, our study presents one of the largest cohorts of patients with anomalous origin of the RCA from the left sinus of Valsalva, and the study stands out for its long-term clinical follow-up.

In theory, a coronary vessel that originates from the contralateral sinus may follow four courses: posterior (retroaortic), septal (subpulmonary), anterior (pre-pulmonary), and interarterial. In practical terms, the risk of ischemia and sudden death is present when the course is interarterial due to a number of mechanisms that can lead to stenosis of the segment, including hypoplasia of the aortic intramural portion and lateral compression, in addition to aortic distensibility and pressure changes. The clinical manifestation is sudden death, typically at young age and after extreme physical exercise, or there is no characteristic presentation. In our case, most patients were asymptomatic for a long time, and the onset of atypical chest pain at adult age was the most common cause for which they were referred for coronary angiography or computed tomography angiography (CTA), rendering the diagnosis.⁵ The mean age of our patients and the clinical manifestations found are consistent with the second group.

When an anomalous origin of the RCA from the left sinus of Valsalva is identified, the evaluation includes a stress myocardial scintigraphy. Although the result is generally negative, this method is important for assessing exercise-induced ischemia. Coronary angiography is indicated to rule out concomitant obstructive coronary disease.⁵

There are three treatment options for patients with anomalous origin of the RCA from the left sinus of Valsalva: medical treatment/observation, coronary angioplasty with stenting, or surgical correction. An intervention may be justified in some cases to prevent sudden death and improve quality of life, but in a significant number of cases it is not necessary.⁹ It is worth noting the high failure rate observed in coronary angioplasty. This is mainly due to difficult selective catheterization and coaxial alignment of the catheter in the RCA, which most often presents an acute angle outlet turned downwards and to the right (slotted hole). The support to introduce the guidewire, balloons, and stents into the coronary artery is therefore impaired, generating more difficulty in successfully concluding the procedure.

The mortality rate for this group in late follow-up is low – similar to that in the general population. These findings, however, may represent selection bias, considering that the most severe manifestation (sudden death) is mainly observed in young adults.⁵

CONCLUSIONS

Anomalous origin of the right coronary artery from the left sinus of Valsalva was an uncommon finding in patients undergoing coronary angiography. Most revascularization procedures were performed due to concomitant atherosclerotic involvement of the vessel, and the mortality rate in late follow-up was low and similar to that in the general population.

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CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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