

## Coronary artery disease in congenital single coronary artery in adults: A Dutch case series

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### Abstract

**AIM:** To assess the current diagnostic and therapeutic management and the clinical implications of congenital single coronary artery (SCA) in adults.

**METHODS:** We identified 15 patients with a SCA detected from four Dutch angiography centers in the period between 2010 and 2013. Symptomatic patients who underwent routine diagnostic coronary angiography (CAG) for suspected coronary artery disease and who incidentally were found to have isolated SCA were analyzed.

**RESULTS:** Fifteen (7 females) with a mean age of  $58.5 \pm 13.78$  years (range 43-86) had a SCA. Conventional

CAG demonstrated congenital isolated SCA originating as a single ostium from the right sinus of Valsalva in 6 patients and originating from the left in 9 patients. Minimal to moderate coronary atherosclerotic changes were found in 4, and severe stenotic lesions in another 4 patients. Seven patients were free of coronary atherosclerosis. Runs of non-sustained ventricular tachycardia were documented in 2 patients, one of whom demonstrated transmural ischemic changes on presentation. Myocardial perfusion scintigraphic evidence of transmural myocardial ischemia was found in 1 patient due to kinking and squeezing of the SCA with an interarterial course between the aorta and pulmonary artery. Multi-slice computed tomography (MSCT) was helpful to delineate the course of the anomalous artery relative to the aorta and pulmonary artery. Percutaneous coronary intervention was successfully performed in 3 patients. Eight patients were managed medically. Arterial bypass graft was performed in 4 patients with the squeezed SCA.

**CONCLUSION:** SCA may be associated with transient transmural myocardial ischemia and aborted sudden death in the absence of coronary atherosclerosis. The availability and sophistication of MSCT facilitates the delineation of the course of a SCA. We present a Dutch case series and review of the literature.

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**Key words:** Congenital heart disease; Coronary artery anomaly; Coronary angiography; Single coronary artery; Coronary artery disease; Multi-slice computed tomography

**Core tip:** A Dutch case series of 15 adult patients with congenital isolated single coronary artery (SCA) are presented. Conventional coronary angiography demon-

strated congenital isolated SCA originating as a single ostium from the right sinus of Valsalva in 6 patients and originating from the left in 9 patients. SCA may be associated with symptomatic transient transmural myocardial ischemia, non-sustained ventricular tachycardia, and aborted sudden death in the absence or presence of coronary atherosclerosis. The availability of multi-slice computed tomography (MSCT) and cardiovascular magnetic resonance imaging facilitates the delineation of the course of the anomalous vessel. MSCT was helpful to delineate the course of the anomalous artery relative to the aorta and pulmonary artery. Percutaneous coronary intervention was successfully performed in 3 patients. Eight patients were managed medically. Arterial bypass graft was performed in 4 patients with the squeezed SCA. The literature addressing SCA is reviewed.

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## INTRODUCTION

A single coronary artery (SCA) is defined as a single aortic orifice or origin providing for all of the coronary blood perfusion of the entire myocardium<sup>[1-3]</sup>. In 1967, Halperin *et al*<sup>[4]</sup> reported the first ante mortem angiographic diagnosis of SCA arising from the left sinus of Valsalva (LSV). SCA is a rare congenital anomaly and occurs as an incidental finding in approximately 0.066% of the coronary angiography (CAG) population<sup>[5]</sup>. SCA has been reported in association with and without atherosclerotic changes<sup>[6,7]</sup> or in association with coronary artery fistulas<sup>[8,9]</sup>, bicuspid aortic valves, and with hypertrophic cardiomyopathy<sup>[1,7,10,11]</sup>.

An equal distribution is found between SCA originating from the right sinus of Valsalva (RSV) and the LSV<sup>[2,12]</sup>. Exact delineation of the course of the abnormal coronaries relative to the aorta and pulmonary artery is of major importance as myocardial ischemia during exertion can be caused by kinking or squeezing of the branches of the anomalous SCA between the aorta and pulmonary artery. CAG is the first diagnostic tool in the detection of a SCA. Once abnormal coronary arteries are suspected, multi-slice computed tomography (MSCT) and cardiac magnetic resonance (CMR) imaging<sup>[13]</sup> scans are excellent tools for non-invasive determination of the course of the abnormal coronaries relative to the aorta and pulmonary artery<sup>[14]</sup>. Determination of the course of incidentally found congenital coronary anomalies during routine CAG without the direct availability of CMR or MSCT scanning is challenging.

We discuss the clinical presentation and angiographic findings of 15 adult symptomatic patients with congenital

isolated SCA incidentally found during routine CAG.

## MATERIALS AND METHODS

Between 2010 and 2013, 15 adult patients with a mean age of  $58.5 \pm 13.78$  years (range 43-86) were diagnosed with a SCA during CAG in 4 Dutch angiography centers (Hospital Group Twente, Almelo; St. Lucas Andreas Hospital, Amsterdam; St. Anna Hospital, Geldrop; Hospital Group Twente, Hengelo; and Gerle Hospital, Zutphen). Indications for CAG were angina pectoris, dyspnea, and syncope.

The angiograms were reviewed by at least two experienced cardiologists who reached a consensus on the origin and course of the SCA. The angiographic variations and the course of the anomalous artery were defined according to the classification of Lipton *et al*<sup>[1]</sup>. The definition of SCA was adopted from Angelini *et al*<sup>[15]</sup> and defined as an isolated coronary artery arising from the sinus of Valsalva through a single ostium and with no evidence of a second ostium, thus being responsible for supplying blood to the entire myocardial tissue, regardless of its distribution.

Significant atherosclerosis was defined as luminal narrowing of  $\geq 75\%$  detected in a main branch of the epicardial coronary arteries. Patients were categorized as having significant single, double, or triple vessel disease when a significant lesion was found in one or more coronary artery branches arising from the SCA and supplying the right coronary artery (RCA), circumflex (Cx), or left anterior descending coronary artery regions. A 12-lead ECG was performed in all patients.

An exercise tolerance test (ETT) was performed in 10 patients, myocardial perfusion test [methoxy-isobutylisonitrile (MIBI) scan] in five patients, and <sup>13</sup>ammonia-adenosine positron emission tomography (positron emission tomography-computed tomography) scan in one patient. MSCT was performed in 6 patients using a retrospective ECG-gated procedure (128-slice, Philips Medical Systems, Best, The Netherlands).

## RESULTS

Patients comprised 8 males and 7 females, aged between 43 and 86 years (mean  $58.5 \pm 13.78$ ). Effort angina pectoris was found in 6 patients, 4 had dyspnea on exertion, 4 complained of atypical chest pain, fainting and pre-syncope, 1 had recurrent syncopal attacks, and 2 presented with acute coronary syndrome. Patients' characteristics are presented in Table 1. Between 2010 and 2013, 8917 coronary angiograms were performed in the 4 Dutch angiography centers all together, with an incidence of 15/8917 (0.017%).

On the resting ECG, all patients were in sinus rhythm and had normal PR intervals. Two patients (patients 3 and 15) had a complete left bundle branch block. One patient (patient 5) had inverted T-waves in the inferior leads. In another patient (patient 8), ECG evidence of an old infero postero lateral infarction was shown. Short

Table 1 Patients' characteristics

Case /gender/age	Clinical presentation	Rest ECG	Risk factors	ETT	MIBI scan	CAD	Management	CAG <sup>1</sup> classification	MSCT
1/F/45	AP, DOE	SR	-	Inconclusive	NA	None	CMM	R-II P	NA
2/M/56	DOE	SR	+	Positive	NA	Intermediate lesion	CMM	R- I	Overestimation of the Cx-lesion
3/F/60	AP	RD SR LBBB	-	Ischemia IL NA	NA	FFR 0.93 Mild	CMM	R-III	NA
4/M/86	ACS	SR	+	Negative	NA	Significant	PCI	L- I	NA
5/M/63	Effort AP	SR Negative T Inferior leads	+	Positive	NA	Significant	PCI	L- II A	NA
6/F/43	ACP, fainting and pre-syncope	SR	+	Inconclusive	Positive <sup>15</sup> N-adenosine PET-CT: normal	None	CABG	L- II B	Course: between aorta and pulmonary artery
7/M/48	AP, syncope	SR NSVT (5 beats)	+	Negative	Negative	None Ergonovine test: No spasm	CMM	L- I	NA
8/F/53	DOE, palpitation	SR RD NSVT (20 beats)	+	NA	Positive	Intermediate lesion	CMM	R- II A	NA
9/M/46	AP, palpitation	SR	-	NA	NA	None	CMM	R- II A	NA
10/M/63	AP	SR	+	Positive	NA	Significant	CABG	L- II B	Course: between aorta and pulmonary artery
11/F/83	NSTEMI	SR	+	NA	NA	Significant	PCI	R-III	NA
12/F/47	ACP	SR	+	Negative	NA	None	CMM	L- II A	NA
13/F/53	CP syncope	SR	-	Negative	Negative	None	CABG	L- II B	Course: between aorta and pulmonary artery
14/M/72	DOE	SR	+	NA	NA	Intermediate lesion	CABG	L- II B	Course: between aorta and pulmonary artery
15/M/41	ACP	SR LBBB	+	Negative	Negative	None	CMM	L- II A	Benign course

<sup>1</sup>Classification according to Lipton *et al*<sup>[1]</sup>. A: Anterior; AP: Angina pectoris; ACP: Atypical chest pain; ACS: Acute coronary syndrome; B: Between aorta and pulmonary artery; CAD: Coronary artery disease; CMM: Conservative medical management; ETT: Exercise tolerance test; F: Female; FFR: Fractional flow reserve; M: Male; CABG: Coronary artery bypass grafting; DOE: Dyspnoea on exertion; N: Normal; NA: Not available; P: Posterior to the aorta; PCI: Percutaneous coronary intervention; R: Right; L: Left; LBBB: Left bundle branch block; MSCT: Multi-slice computed tomography; SR: Sinus rhythm; -: Absent; +: Present (hypertension, smoking, obesity, hypercholesterolemia); PET-CT: Positron emission tomography-computed tomography; NSVT: Non-sustained ventricular tachycardia; RD: Repolarization disturbances; Cx: Circumflex; CAG: Coronary angiography; NSTEMI: Non-ST elevation myocardial infarction; MIBI: Methoxy-isobutyl-isonitrite; CP: Chest pain.

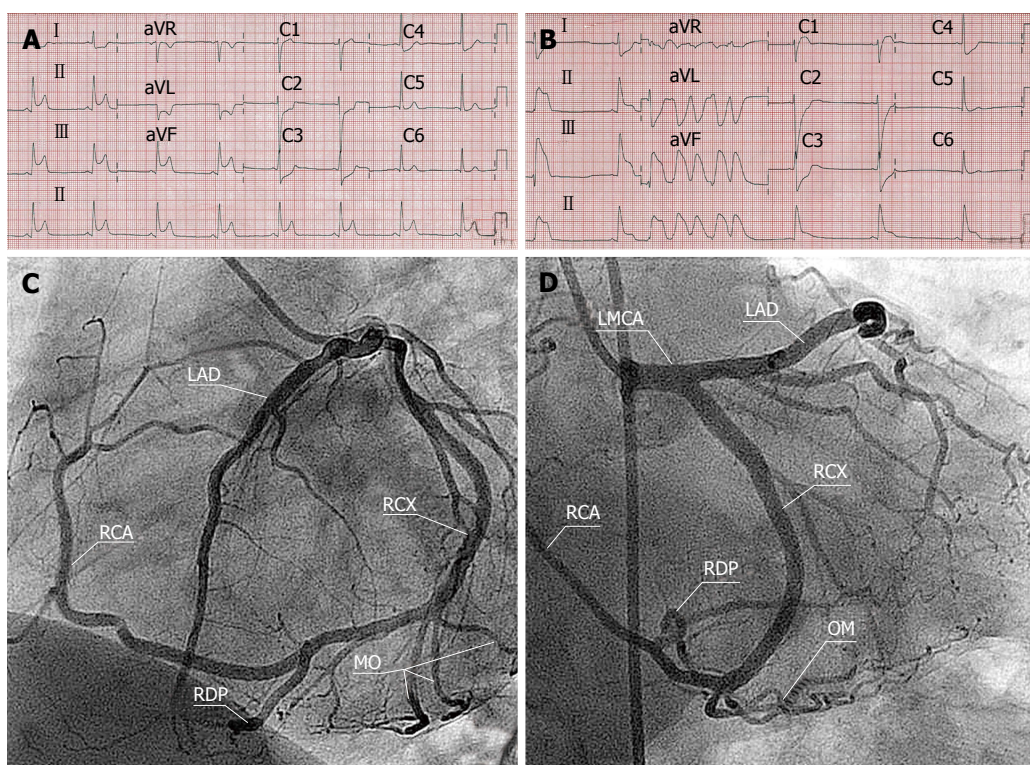
runs of non-sustained ventricular tachycardia (NSVT), varying from 5 to 20 beats/min with a frequency of 176/min and duration of 1800 ms, were documented in 2 patients (patients 7 and 8) (Figure 1).

ETT was inconclusive in 2 patients (1 and 6). Diagnostic CAG showed no significant coronary artery lesions in either patient. In patient number 6, the PET scan was positive due to kinking and squeezing of the SCA with a course between the aorta and pulmonary artery. This patient underwent coronary artery bypass grafting (CABG) whereby a mammary arterial graft was anastomosed to the RCA. In 3 patients (2, 5 and 10), the ETT was positive for myocardial ischemia. Of the 3 patients with positive ETT, 1 had significant CAD and underwent percutaneous coronary intervention (PCI). The other 2 patients demonstrated an intermediate lesion distally located in the coronary arterial tree and were managed medically. The ETT was negative in 5 patients (4, 7, 12, 13 and 15). Despite a negative ETT, patient number 4 showed a significant coronary lesion on CAG and underwent PCI. Pa-

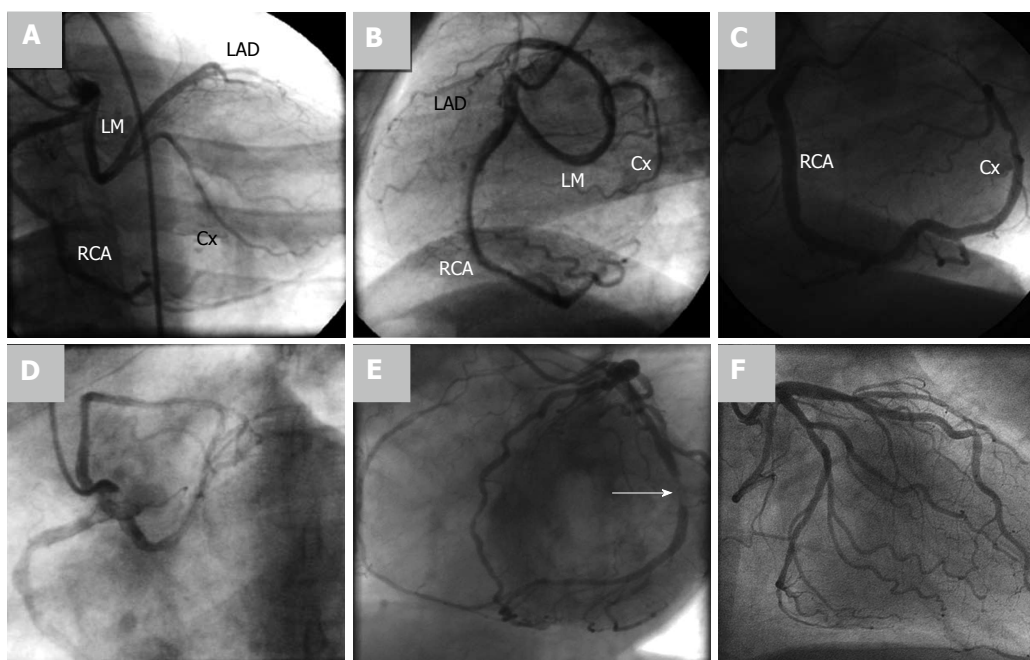
tient number 7 had no significant coronary artery lesions and the ergonovine test was negative. Patient number 8 had a positive MIBI scan and CAG showed an intermediate lesion, which was managed medically. MSCT scan of 5 patients (6, 10, 13, 14 and 15) demonstrated an interarterial course and they underwent CABG, whereby a mammary arterial graft was anastomosed to the RCA in 4 and the fifth showed a benign course.

Conventional CAG demonstrated a SCA originating as a single ostium from the RSV in 6 patients and SCA originating as a single ostium from the LSV in 9 patients (Figures 2 and 3).

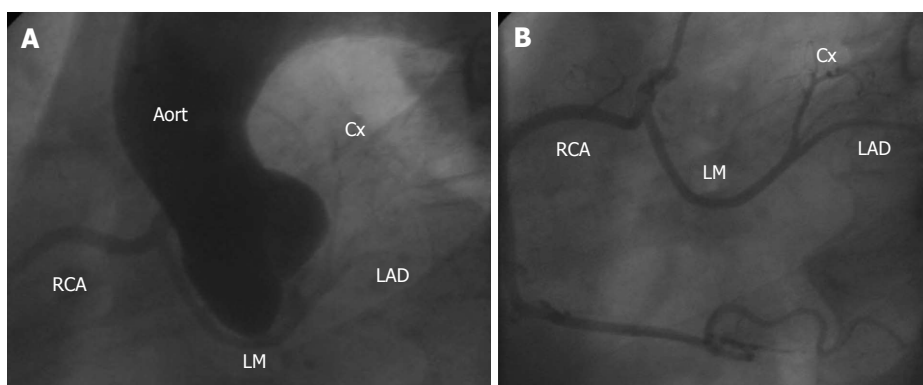
MSCT was performed in 6 patients (2, 6, 10, 13, 14 and 15). In patient 2, MSCT confirmed the diagnosis of SCA but gave an overestimation of the severity of the coronary lesion in the Cx trajectory, which was not significant (0.93) by fractional flow reserve measurement (Figure 4). In 4 patients (6, 10, 13 and 14), SCA was also proven by MSCT depicting clearly the course of the SCA running between the aorta and the pulmonary artery



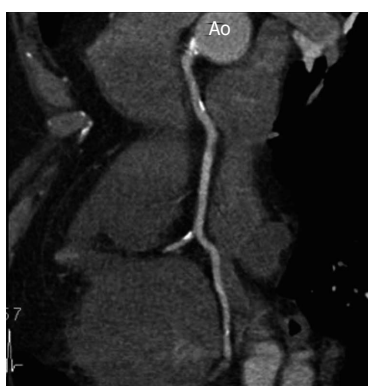
**Figure 1 Resting electrocardiograph.** A: Electrocardiograph during chest pain depicting transmural ischemia in the infero-posterior leads; B: Followed by a non-sustained monomorphic ventricular tachycardia; C: Coronary angiography showed absence of the right coronary ostium and a single coronary artery arising from the left sinus of Valsalva with normal origin of the left coronary artery (LCA) having normal anatomical course of the left main stem, the left anterior descending, and the circumflex artery (Lipton L- I ); D: The LCA supplies the entire myocardial tissue. No significant stenoses were found. The right coronary artery (RCA) appeared as a continuation of the distal left circumflex artery to the right atrioventricular groove and terminated near the RSV (Lipton L- I ). LAD: Left anterior descending; LMCA: Left main coronary artery; RCX: Ramus circumflexus; RDP: Ramus descending posterior; OM: Obtuse marginal.



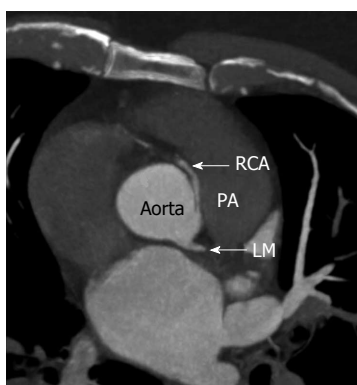
**Figure 2 Coronary angiography frame.** A: Coronary angiography frame of right anterior oblique projection with cranial angulation; B: Left lateral (LL) projection showing a single origin of the right and left coronary arteries from a common right coronary ostium (Lipton R- II P), the long curved left main stem and right dominance are delineated; C: Coronary angiography frame in LL projection demonstrating a single coronary artery originating from the right sinus of Valsalva (RSV) giving the left anterior descending (LAD) and continued as the circumflex artery (Lipton R- I ); D: Coronary angiography frame in left anterior oblique view demonstrating a single coronary artery arising from RSV as a single unique ostium (Lipton R-III); E: Coronary angiography frame in left anterior oblique view showing a single coronary artery originating from the left sinus of Valsalva. The terminal branch of circumflex artery represented the right coronary artery (Lipton L- I ). Significant stenosis of the mid circumflex artery is demonstrated (white arrow); F: Coronary angiography frame demonstrates appearance of both right and left coronary arteries on injection of left sinus of Valsalva, as a single common ostium (Lipton L- II A). Cx: Circumflex artery; RCA: Right coronary artery.



**Figure 3 Angiography.** A: Supravulvar aortogram in left anterior oblique projection illustrating a single origin of the coronary arteries originating from the right sinus of Valsalva (Lipton R- II A); B: Selective coronary angiography frame in left anterior oblique view showing a single coronary artery from the right sinus of Valsalva. Cx: Circumflex artery; LAD: Left anterior descending; RCA: Right coronary artery.



**Figure 4** Transverse Multi-slice computed tomography scan in subtype (Lipton R- I) demonstrating the origin of the single coronary artery arising from the right sinus of Valsalva supplying the whole heart.



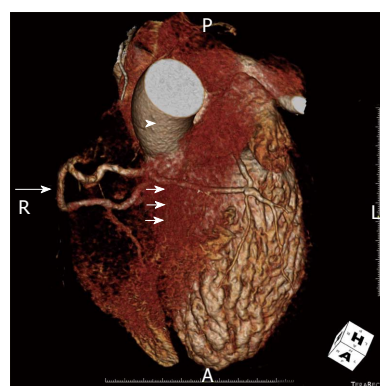
**Figure 5** Volume-rendered image in subtype (Lipton L- II B) demonstrating the inter-arterial course of the right coronary artery between the aorta and pulmonary artery.

(Figures 5, 6). MSCT in patient 15 demonstrated a benign course of the SCA (Figure 7).

## DISCUSSION

### Historical background of classifications

The coronary arterial circulation may rarely be supplied by a SCA arising from either the right, left or posterior



**Figure 6** Three-dimensional volume-rendered image in subtype (Lipton L- II B) demonstrating the inter-arterial course of the right coronary artery (long arrow) between the aorta (arrowhead) and semitransparent pulmonary artery (short arrows).

sinus of Valsalva<sup>[16]</sup>. The course of the SCA can be highly variable. In the last century, different classification systems for SCA based on necropsy findings and angiographic variants were suggested in the fifties by Smith<sup>[3]</sup> (3 types), in the seventies by Lipton *et al*<sup>[1]</sup>, in the eighties by Roberts<sup>[17]</sup>, and finally through the nineties by Shirani *et al*<sup>[2]</sup> and Roberts *et al*<sup>[18]</sup>.

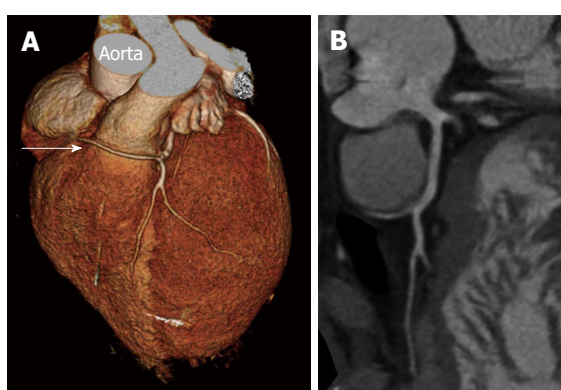
Recently, a clinically useful classification scheme has been published, using either subgroups based on the site of origin and course of the anomalous coronary artery or descriptive anatomic terminology. In 2005, Rigatelli *et al*<sup>[19,20]</sup> based his classification on clinical significance of the anomaly and launched a global practical classification of four categories (class A: benign; class B: relevant due to fixed myocardial ischemia; class C: severe, involved in sudden cardiac death (SCD); and class D: critical due to worsened clinical picture). The clinical significance and management of the various types of SCA are different as shown in Table 2.

Cheitlin *et al*<sup>[21]</sup> expressed the pathological significance of a SCA or of both coronary arteries originating from the RSV when the anomalous artery that supplies the left coronary distribution passes leftward with an inter-arterial course between the aorta and pulmonary trunk, rendering

**Table 2** Clinical-significance-based classification of coronary artery anomalies by Rigatelli *et al.*<sup>[19,20]</sup>

Class	Subtypes	Clinical significance	Current series
A	E.g., ectopic origin of Cx from RSV <sup>1</sup>	Benign natural history, asymptomatic careful follow-up with conservative medical management or percutaneous intervention	Patients: none <sup>1</sup>
B	Ectopic origin of Cx from the RCA R- I , R- II , R- III anterior/posterior course <sup>2</sup>	Relevant, related to myocardial ischemia Careful follow-up with conservative medical management or percutaneous intervention	Patients: 1, 2, 3, 7, 8, 9, 12, 15
C	L- I , L- II , L- III anterior/posterior course <sup>2</sup> R- I , R- II , R- III between/interseptal course <sup>2</sup>	Severe, potentially related to sudden cardiac death Requires surgical treatment	Patients: 6, 10, 13, 14
D	L- I , L- II , L- III between/interseptal course <sup>2</sup>  B or C subgroups with concomitant coronary atherosclerosis	Critical, class B or C with superimposed coronary artery atherosclerotic disease Requires urgent percutaneous management or surgical treatment	Patients: 4, 5, 11

<sup>1</sup>Not included in the current paper; <sup>2</sup>Classification according to Lipton *et al.*<sup>[11]</sup>. Cx: Circumflex coronary artery; L: Left; R: Right; RCA: Right coronary artery; RSV: Right sinus of Valsalva.



**Figure 7** Single coronary artery. A: Three-dimensional volume-rendered image of benign course of right coronary artery (arrow) from left sinus of Valsalva (Lipton L-IIA); B: Transverse multi-slice computed tomography scan in subtype (Lipton L-IIA) demonstrating the origin of the single coronary artery arising from the left sinus of Valsalva supplying the whole heart.

it prone to compression and kinking on physical exercise. This variant is considered malignant since it is associated with SCD in adolescents and young adults, especially on the athletic arena. It has been found that anomalous origin of the left coronary artery from the right aortic sinus is consistently related to sudden death in more than half of the cases (53%)<sup>[18]</sup>.

On the other hand, when a SCA originating from the LSV or both coronary arteries arising from separate ostia located in the LSV with the RCA passing inter-arterially (between the aorta and pulmonary trunk, e.g., subtype Lipton L- IIB) is less deleterious even though compression can occur but SCD is a rare event. Four of our 15 patients (patients 6, 10, 13 and 14), having the above-mentioned subtype, underwent successful arterial bypass grafting to the RCA.

In a necropsy series, SCA was found in 18% of subjects. Fifty percent arose from the RSV and 50% originated from the LSV. Sudden death was twofold more frequently associated with the SCA arising from the RSV (18%) compared with those from the LSV (9%)<sup>[12]</sup>.

Coronary artery anomalies are associated with life threatening symptoms and may cause SCD during or

after strenuous exercise. The most common congenital coronary artery anomalies causing SCD involve an anomalous origin of either the right or left coronary artery arising from the left or the RSV, respectively<sup>[22]</sup>. SCD is common (82%) when the anomalous LCA has an inter-arterial course passing between the aorta and main pulmonary artery<sup>[12]</sup>. Moreover, SCD may rarely occur after surgical repair<sup>[23]</sup>. The incidence is very low and estimated at 0.024% to 0.098% in the general population<sup>[1,5,9,19,24]</sup>. The incidence of all coronary artery anomalies in the necropsy series is approximately 0.23% and varying from 0.3% to 13% in the angiographic series<sup>[1,9,25,26]</sup>. Recently, the incidence of SCA, using dual-source computer tomography angiography, was estimated at 0.05% in the Chinese adult population<sup>[27]</sup>. SCA may be associated with longevity and has been reported in an octogenarian<sup>[28]</sup>.

**Diagnostic modalities:** The correct diagnosis of a SCA and its course is not always easily made based on conventional CAG only. Precise delineation of anatomical and functional characteristics requires further complementary diagnostic modalities such as MSCT or CMR<sup>[29-31]</sup>.

**Conventional CAG:** Isolated SCA may be incidentally detected on routine CAG<sup>[32]</sup>, as was the case in our current series. Even with multiple projections and different angiographic views and the use of a pulmonary artery catheter, the identification of the origin and proximal course of the vessel can be difficult<sup>[13]</sup>. Serota *et al.*<sup>[33]</sup> proposed an angiographic technique (the dot-and-eye method) for rapid identification of the course of SCA but even with this method, identification remains difficult.

**MSCT CAG:** MSCT has been very useful in the diagnosis and identification of the origin and course of SCA<sup>[28,32]</sup>. Although the radiation dose using new algorithms is decreasing, this rapidly developing non-invasive technique still has the disadvantage of radiation exposure. However, the spatial resolution (0.4-0.6 mm<sup>3</sup>) is higher than CMR and the temporal resolution of 64-slice double source MSCT is around 83 ms<sup>[30,31,34-37]</sup>. In 5 of our patients (patients 6, 10, 13, 14 and 15) of the current series, 128-slice

MSCT confirmed the diagnosis of a SCA with clear demonstration of the inter-arterial course of the RCA originating from the LSV in four (Figure 6) and a benign course of the RCA from LSV in one (Figure 7).

**Cardiovascular MR imaging:** This technique has the advantage of not using ionizing radiation and has no need for the use of iodinated nephrotoxic ionic or non-ionic contrast agents. Image acquisition occurs with fairly good spatial and temporal resolution, but acquisition and imaging time is long, which makes routine use difficult and time consuming. Cardiovascular magnetic resonance proved to be useful in determining the anatomy and functional significance of SCA<sup>[38]</sup>. Both the MSCT and the CMR imaging techniques have the additional advantage of 3-D reconstruction of the areas of the coronaries relative to the aorta and pulmonary artery. This makes a definitive diagnosis of squeezed aberrant coronary arteries between the great vessels feasible<sup>[13]</sup>.

### Treatment

The detection of atherosclerotic coronary artery disease (CAD) in the presence of coronary anomalies is of practical importance, especially when a decision between PCI and CABG has to be made. For diagnostic and therapeutic reasons, the knowledge of possible variations of the coronary anatomy, their different origin, and their course is of pivotal importance. Symptomatic patients with associated significant CAD may be treated with routine interventions such as PCI or CABG<sup>[6,39]</sup>. Angiographic recognition of coronary artery anomalies prior to surgery is of great importance. During operation, surgical complications may occur if an unrecognized anomalous vessel is excluded from perfusion during cardiopulmonary bypass or if the surgeon inadvertently damages an artery with an anomalous pathway.

Because of the reported high mortality, the occurrence of “symptomatic or asymptomatic” squeezing of SCA, regardless of the degree of atherosclerosis or site of origin, justifies arterial grafting, as was shown in 4 of our series (patients 6, 10, 13 and 14).

Significant atherosclerotic CAD<sup>[37]</sup> in association with coronary artery anomalies has been reported in 26%-60% of cases<sup>[1,2,40-42]</sup>. Rigatelli *et al.*<sup>[43]</sup> suggested that benign coronary artery anomalies are not associated with or involved in the development of premature atherosclerotic CAD. Indeed the high percentage of coronary artery stenosis could be biased by the indication to perform CAG as SCA is mainly found during this diagnostic procedure. Only 4 of our 15 (27%) patients (patients 4, 5, 10 and 11) had significant CAD and 3 of them required percutaneous intervention. When the SCA does not course between the aorta and pulmonary artery, it is not vulnerable to acute angulations or kinking of the coronaries. SCA may be associated with longevity and patients in the 7<sup>th</sup> and 8<sup>th</sup> decade of life have been reported<sup>[3,12,13,44-47]</sup>, as was the case in 2 octogenarians from our current series (patients 4 and 11).

Although a SCA is often a benign congenital anomaly,

in which sudden death is a rare complication, different diagnostic modalities should be used to exclude an inter-arterial course between the aorta and pulmonary artery to detect patients at risk for serious complications.

Congenital coronary artery anomalies, detected at necropsy, associated with sudden death and without antecedent signs have been recognized in calves<sup>[48]</sup>. SCA is not limited to the human race, it has also been reported in other mammals such as horses<sup>[49]</sup>, syrian hamsters<sup>[50]</sup> and minipigs<sup>[51]</sup>.

As was shown in our patient's population, SCA can be associated with longevity. It has been documented up till the 8<sup>th</sup> decade of life. In the adult population, SCA-isolated or in association with acquired atherosclerotic changes-may cause severe sequelae. In some cases without CAD, the course of the SCA may be malignant.

SCA may be associated with symptomatic transient transmural myocardial ischemia, NSVT, and aborted sudden death in the absence or presence of coronary atherosclerosis. The availability of MSCT and CMR facilitates the delineation of the course of the anomalous vessel. The accurate delineation of the course of the anomalous vessel is of great importance even in patients without CAD and in cases of surgical intervention where anatomic details of the course of the vessel are of importance.

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## COMMENTS

### Background

Single coronary artery (SCA) is a rare congenital anomaly and occurs as an incidental finding in approximately 0.066% of the coronary angiography (CAG) population. SCA has been reported in association with and without atherosclerotic changes or in association with coronary artery fistulas, bicuspid aortic valves, and with hypertrophic cardiomyopathy.

### Research frontiers

CAG is the first diagnostic tool in the detection of a SCA. Once abnormal coronary arteries are suspected, multi-slice computed tomography (MSCT) and cardiac magnetic resonance (CMR) imaging scans are excellent tools for non-invasive determination of the course of the abnormal coronaries relative to the aorta and pulmonary artery. Determination of the course of incidentally found congenital coronary anomalies during routine CAG without the direct availability of CMR or MSCT scanning is challenging.

### Innovations and breakthroughs

Percutaneous coronary intervention was successfully performed in 3 patients. Eight patients were managed medically. Arterial bypass graft was performed in 4 patients with the squeezed SCA. The literature addressing SCA is reviewed.

## Applications

Congenital coronary artery anomalies, detected at necropsy, associated with sudden death and without antecedent signs have been recognized in calves. SCA is not limited to the human race, it has also been reported in other mammals such as horses, syrian hamsters and minipigs.

## Peer review

This paper showed that the availability and sophistications of MSCT facilitated the delineation of the course of a SCA. The authors presented a Dutch case series and review of the literature. This is an interesting report for clinical practice. Overall the report appears to be carefully examined and data adequately discussed.

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