Case Report

Percutaneous Coronary Intervention and Stenting in a Single Coronary Artery Originating from the Right Sinus of Valsalva

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Key words: **Single coronary artery, percutaneous intervention, dot and eye method.**

Percutaneous revascularisation in anomalous coronary arteries poses certain technical difficulties, especially in the selective cannulation of the diseased vessel and the coaxial positioning of the guiding catheter for optimum stability. We report a case of successful stent implantation in a single coronary artery (SCA) arising from the right sinus of Valsalva (SoV). The ART 4 (Boston Scientific) guiding catheter provided excellent back-up support for stent delivery in the diseased circumflex artery. We also discuss the anatomical variations of an SCA originating from the right SoV, their angiographic recognition, the clinical implications, as well as technical considerations that should be taken into account when performing percutaneous interventions in these vessels.

Manuscript received:
July 8, 2011;
Accepted:
March 12, 2012.

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The incidence of coronary artery anomalies in adult angiographic series ranges between 0.2-1.3%;[1-5] however, the incidence of these anomalies in the general population may be underestimated, since many individuals are asymptomatic. Most of these anomalies are clinically benign, but some are associated with a high risk of adverse cardiac events, even in the absence of atherosclerosis.[6-9] The specific origin and course of the anomalous vessel are the factors that determine an adverse or benign outcome. Coronary arteries originating from a single coronary ostium in the aorta are extremely rare. The prevalence of a single coronary artery (SCA) is reported to be less than 3% of all major coronary artery anomalies.[10] In this report, we describe a case of anomalous origin of the left coronary artery (LCA) from the right sinus of Valsalva (SoV). PCI and direct implantation of a new-generation, drug-eluting stent was performed in the circumflex artery (Cx), which had a severe lesion in its mid-segment. We also present a CT angiography image demonstrating that the left main (LM) coronary artery had an intraseptal course before its bifurcation to the left anterior descending (LAD) and Cx arteries.

Case presentation

A 55-year-old man with episodes of chest pain of suspected cardiac cause and a positive exercise treadmill test at moderate workload was referred for coronary angiography. His past medical history included hypertension, dyslipidaemia and a 40 pack-year history of cigarette smoking. He denied any previous episodes of dizziness or syncope. Physical examination was unremarkable. Routine 12-lead ECG, chest X-ray and echocardiogram showed no pathological findings.

Cardiac catheterisation was performed through the right femoral approach, but we

were unable to cannulate the LCA, despite repeated attempts with different catheters. Non-selective injection of contrast in the left coronary sinus failed to demonstrate a left-sided coronary origin. Selective cannulation of the right coronary artery (RCA) with a 6-Fr JR 4.0 diagnostic catheter revealed that the entire coronary circulation originated from a single ostium within the right SoV (Figure 1). The RCA continued in the right atrioventricular groove to the origins of the posterior descending artery and posterolateral branch and was free of significant atherosclerotic disease. The LM appeared to have an intraseptal course before its bifurcation to the LAD, which had only mild irregularities, and the Cx, which had a severe, discrete stenosis in the middle segment (Figure 2). Left ventriculography was normal, with an estimated ejection fraction of 60%. In order to accurately delineate the course of the abnormal coronary artery, 64-slice CT angiography (GE LightSpeed Multidetector CT Scanner) was performed, which clearly depicted the SCA arising from the right SoV and the LM and passing between the aorta and the right ventricular outflow tract (Figures 3 & 4).

The single coronary ostium was engaged in a coaxial fashion with an ART 4.0 SH guiding catheter (Boston Scientific) and the lesion was crossed with a 0.014" balanced heavy-weight wire (Guidant Corp). Direct stenting of the lesion was successfully performed with a 3.0 × 15 mm everolimus-eluting stent (Promus, Boston Scientific). The final angiographic result was excellent, with no residual stenosis (Figure 5). The patient had an uneventful hospital course and was discharged on dual antiplatelet therapy, statin, and β-blockers. An exercise tolerance test performed 6 months after the procedure did not show any signs of ischaemia.

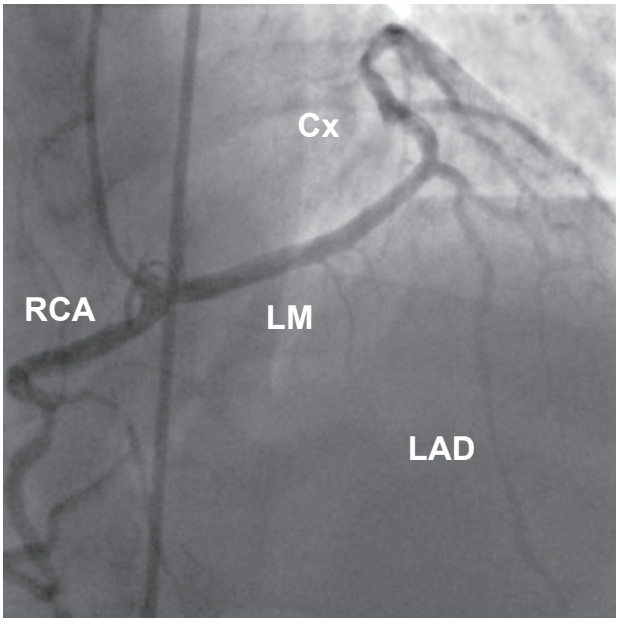

Figure 1. Right anterior oblique 30° - cranial 20° view, showing a single coronary artery originating from the right sinus of Valsalva.

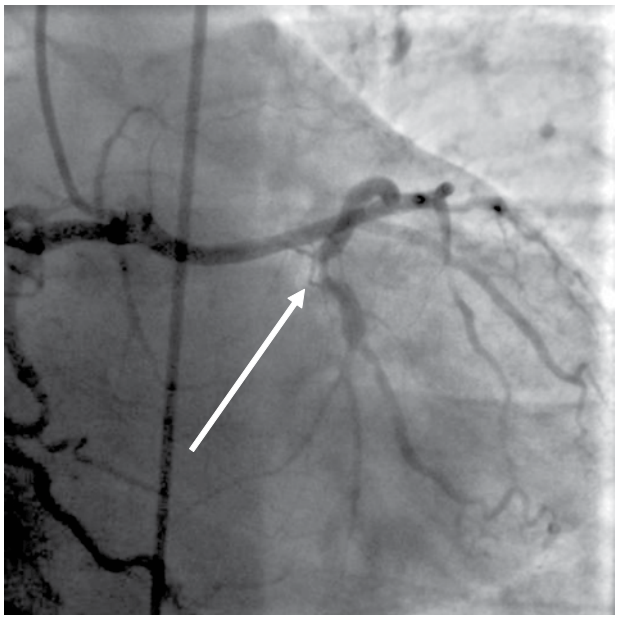

Figure 2. Significant stenosis in the middle segment of the circumflex artery (arrow).

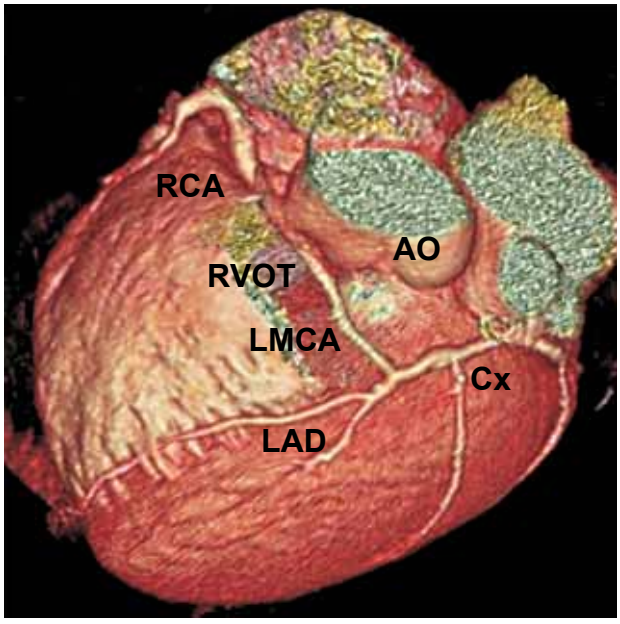

Figure 3. A 3-D volume rendering image where the left main stem (LMCA) is shown to pass in front of the aortic valve, between the right sinus of Valsalva and the right ventricular outflow tract (RVOT).

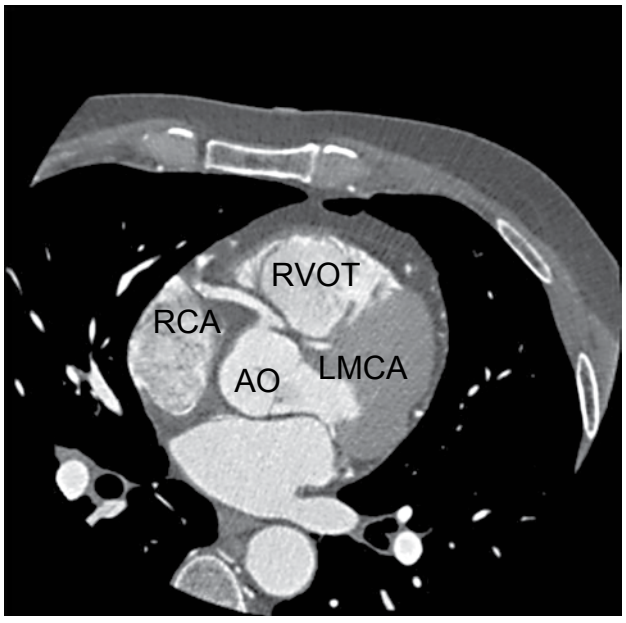

Figure 4. A cross-sectional image, demonstrating the left main coronary (LMCA) artery passing between the aorta (AO) and the right ventricular outflow tract (RVOT).

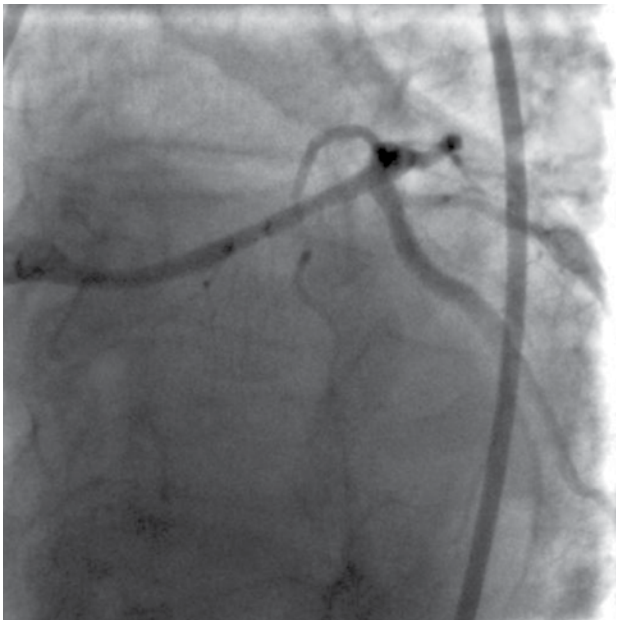

Figure 5. Final angiographic result after stent deployment (left anterior oblique 30°- caudal 20° projection).

Discussion

The SCA refers to the origination of both the left and right coronary arteries from a single aortic ostium and was first reported by Hyrtl in 1841,[29] but the first diagnosis with angiography was made in 1967. In several large series, the incidence of anomalous origin of the LCA from the right SoV as a single coronary artery ranged between 0.019-0.066% and accounted for 1.2-6.1% of all coronary anomalies.[5-7] Yamanaka et al reported an SCA arising from the right SoV in 0.019% of 126,595 patients undergoing coronary angiography.[1] In a study of 142 patients with an SCA, which also included patients with congenital heart disease, the SCA arose from the right SoV in 49% of patients and from the left SoV in 45% of patients. The latter study also included patients with an SCA originating from the pulmonary trunk, and major congenital cardiac malformations were present in 41%. The most common anomalies were transposition of the great vessels, coronary artery fistula, improper division of the truncus arteriosus, tetralogy of Fallot and bicuspid aortic valve. In a series of 4535 adult patients from northern Greece who underwent coronary angiography at our institution, the incidence of SCA originating from the right SoV was 0.044% (2 cases).

Classification

An SCA originating from the right SoV may have a common ostium with the RCA or arise from its proximal segment. According to the relationship of the anomalous left main with the aorta and the pulmonary artery, the SCA can be classified into five anatomical subtypes: 1) septal, 2) interarterial, 3) anterior, 4) posterior, and 5) mixed. Lipton et al proposed a very useful angiographic classification of SCAs,[6] which was further modified in 1990 by Yamanaka and Hobbs, based on ostium location, anatomical distribution and course of the transverse trunk (Table 1). According to this classification, our patient had a type R/II/S (septal) SCA. In this type, which is the commonest variant, the left main runs an intramyocardial course through the ventricular septum along the floor of the right ventricular outflow tract and then surfaces to a normal epicardial position in the mid-septum, where it divides into the LAD and Cx. The Cx then courses toward the aorta, and the LAD appears relatively short.

The most recent classification by Shirani and Roberts (Figure 6) takes into account every anatomical probability, whether it has been reported or not.[7] An ostium originating from the left SoV is type I, and if it arises from the right SoV it is type II. The most common type is II/B. In our patient, according to this classification, the SCA was type II/B/3.

Table 1. Modified Lipton's classification of the types of isolated single coronary artery.

	Code	Description
Ostial location	R	Right sinus of Valsalva
	L	Left sinus of Valsalva
Anatomical distribution	I	The solitary dominant vessel follows the course of either a normal right or left coronary artery (RI or LI)
	II	One coronary artery arises from the proximal part of the normally located other coronary artery (RII or LII)
	III	LAD and Cx arise separately from common trunk originating from the right SoV (RIII)
Course of the transverse trunk	A	Anterior to the great vessels
	B	Between aorta and pulmonary artery
	P	Posterior to the great vessels
	S	Septal type: a part of the route passes through the interventricular septum
	C	Combination of diverse routes

LAD – left anterior descending artery; Cx – circumflex artery; SoV – sinus of Valsalva.

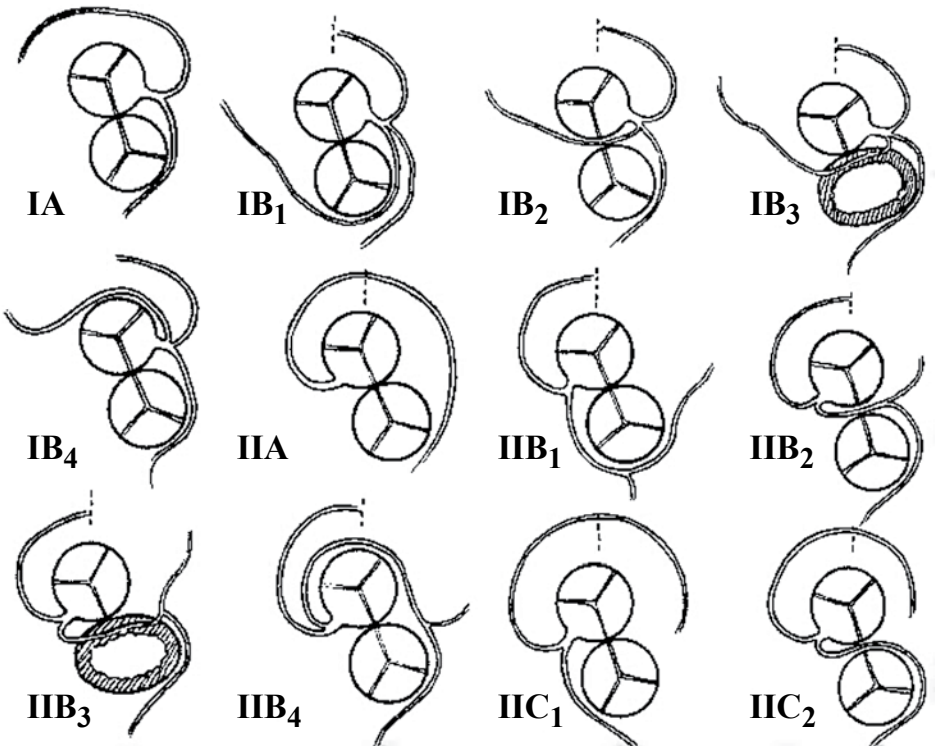

Figure 6. Shirani and Roberts classification. Reprinted from reference #7, with permission from Elsevier.

Clinical implications

Coronary artery anomalies are usually discovered as incidental findings during coronary angiography for suspected coronary artery disease. The widespread use of coronary angiography has resulted in more frequent detection of these anomalies, and their clinical significance is coming to be better appreciated. Certain origins and courses are thought to be associated with an increased risk of myocardial ischaemia, infarction, congestive heart failure, ventricular arrhythmias, syncope or sudden cardiac death (SCD), while others are generally considered benign.[5-12,17]

The pattern most often thought to be associated with increased clinical risk is that of an ectopic coronary artery arising from an inappropriate SoV (i.e. LM from the right SoV or RCA from the left) and passing between the aorta and the main pulmonary artery. Indeed, 80% of autopsies in athletes with SCD and coronary anomalies revealed that the anomalous vessel had a course between the aorta and the pulmonary artery.[11] Moreover, anomalous origin of the coronary arteries

has been reported as the cause of 15-17% of SCDs in athletes, second commonest after hypertrophic cardiomyopathy.[12] In another study, coronary anomalies were responsible for 12% of sports-related SCDs versus 1.2% of non sports-related deaths in 14-40 year-olds, suggesting that coronary anomalies are lethal mostly during or shortly after strenuous physical activity.[28] The pathophysiological mechanism is not exactly clear. Various mechanisms, which may act alone or in combination, have been proposed to explain this, including: direct compression during exercise between the pulmonary artery and aorta when these vessels dilate; inadequate flow through a narrow slit-like orifice of the anomalous vessel, which can collapse during exercise; acute kinking of the long LM and spasm of the proximal portion of the LM.

On the other hand, coronary arteries that pass anterior to the pulmonary trunk, posterior to the aorta, or through the interventricular septum are rarely associated with these complications; thus, patients with these anomalies are usually asymptomatic unless atherosclerotic disease is present. It is therefore important to recognise the presence and the proximal course of the anomalous coronary artery at the time of the angiogram, in order to estimate the clinical risk and plan subsequent management.

"Dot and Eye" method

The course of the anomalous coronary arteries is best assessed in the 30° right anterior oblique (RAO) view with the "Dot and Eye" method,[13] though in some cases this may still be difficult (Figure 7). In the "septal" type, the LM and Cx form an ellipse-like configuration (similar to the appearance of an eye), with the LM forming the inferior and the Cx the superior portion of the ellipse. Septal perforators coming off the LM help to differentiate it from other subtypes. The most potentially fatal variant is the "interarterial" subtype. In the RAO view, a radiopaque dot representing the artery seen end-on is noted on the anterior aspect of the aorta. In the third subtype the LM courses the "anterior" free wall of the right ventricle and the pulmonary artery. Again in the RAO projection, the LM and the initial portion of the Cx will form an ellipse (eye). The LM now forms the upper portion of the ellipse, and the Cx forms the lower portion. In the fourth pattern, the LM passes "posterior" to the aorta and a radiopaque dot is noted on its posterior aspect. Figure 8 shows the typical angiographic "eye" sign in our case (the LM forming the inferior and the Cx the superior portion of the ellipse), which, along with the presence of septal perforators branching off the left main, indicated that the anomalous vessel had an intramyocardial course.

Non-invasive methods to assess coronary anatomy

Numerous studies suggest that computed tomography and magnetic resonance imaging add valuable information to coronary angiography, in the diagnosis and delineation of the origin and course of anomalous coronary arteries.[14-17] The new generation of CT scanners with improved reconstruction algorithms provides better spatial and temporal resolution, and a shorter scan time, permitting better image quality

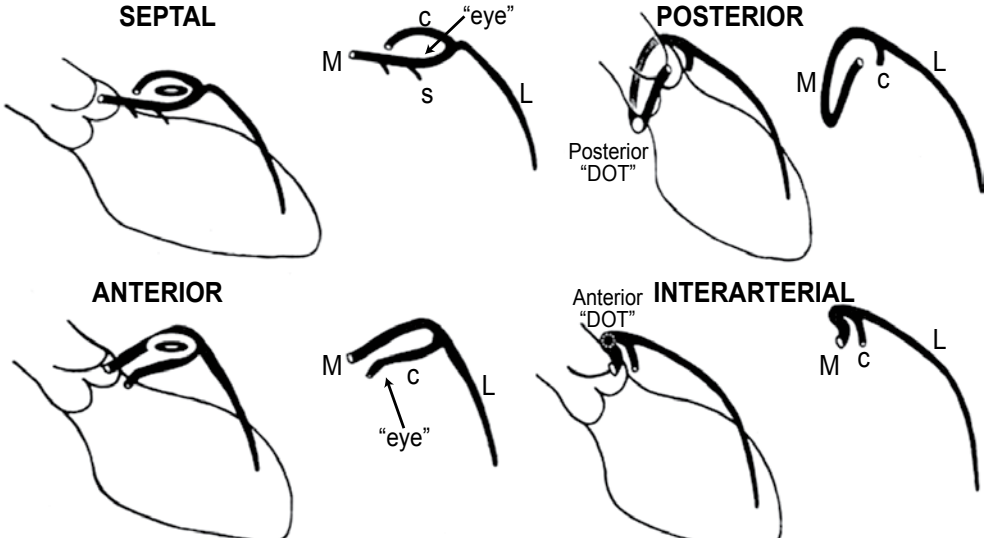

Figure 7. The "Dot and Eye" method in right anterior oblique imaging (Eye: loop formed by the left main (LM) and circumflex (Cx) arteries. Dot: vessel seen end-on). Reprinted from reference #13, with permission from Elsevier.

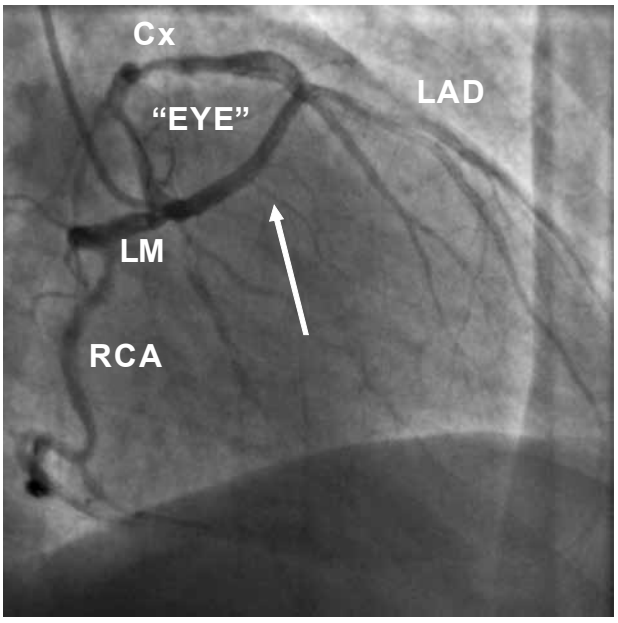

Figure 8. Angiography in 30° right anterior oblique projection. An "eye" formed by the anomalous left main and circumflex arteries is outlined. Septal perforators coming off the left main (arrow).

and reduced motion artefacts. MR angiography has also been shown to be accurate, with the advantage of requiring no iodinated contrast or radiation exposure. The non-invasive nature and the ability to acquire complex 3-dimensional views have made these imaging modalities very useful in evaluating coronary arteries, as well as their relation with surrounding anatomic structures. In our case, multislice CT coronary angiography demonstrated the anomalous LCA passing between the aorta and the right ventricular outflow tract.

Technical considerations

Catheter-based revascularisation in anomalous coronary arteries is a therapeutic challenge for interventional cardiologists. Percutaneous coronary intervention in these patients requires proper angiographic recognition of anatomical details, such as the orifice configuration, the exit angulation from the aorta, the route of the anomalous artery, and the location of the atherosclerotic lesion.[18-25]

The major factor determining successful PCI is the selection of the guiding catheter. Selective cannulation can be difficult and time-consuming; hence, knowledge of the variations in coronary artery origin can help in selecting the appropriate catheters for diagnostic and therapeutic interventions. Stable guiding catheter seating and optimal back-up support is the key to abnormally arising coronaries, in order to aid wire and device passage. Back-up support can be increased by using: a) a larger diameter or a different-shaped guiding catheter, or with deep intubation into the target vessel, b) a stiffer wire or buddy wires, c) a buddy balloon, or d) the anchor technique.[26] Das and Wysham described a double-wire strategy, using a wire passed into the RCA to enhance guiding catheter stability, in order to facilitate the passage of a second wire into an anomalous Cx.[27]

The ART 4 SH guiding catheter and the stiffer BHW guide wire used in our case provided the maximum support required for smooth passage of the stent. The tip of this catheter engaged well in the anomalous artery with its curve resting stably against the opposite aortic wall.

Theoretically, performing PCI in an SCA increases the procedural risk, since a complication leading to dissection (e.g. guiding-catheter induced) of the single ostium may be catastrophic. Practically, since the single ostium usually has a large calibre, the risk of guiding catheter damage is very low.

Conclusion

We report a case with the septal type of anomalous origin of the left coronary artery from the right SoV. Since this variant has been reported to be of relatively benign clinical course and the lesion in the circumflex artery was amenable to angioplasty, we decided to treat this patient percutaneously with direct stent implantation.

PCI in anomalous coronary arteries is a feasible therapeutic strategy; however, accurate topographic identification of the origin and proximal course of the anomalous vessel is of paramount importance before proceeding in these interventions, in order to plan the most appropriate type of medical, percutaneous or surgical treatment. Furthermore, selection of the appropriate guiding catheter and other equipment is essential to the technical success of the procedure.

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