ACCEPTED MANUSCRIPT

Accepted manuscripts are the articles in press that have been peer reviewed and accepted for publication by the Editorial Board of the Vojnosanitetski Pregled. They have not yet been copy edited and/or formatted in the publication house style, and the text could still be changed before final publication.

Although accepted manuscripts do not yet have all bibliographic details available, they can already be cited using the year of online publication and the DOI, as follows: article title, the author(s), publication (year), the DOI.

Please cite this article PRIMARY PCI IN A PATIENT WITH ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE OPPOSITE SINUS OF VALSALVA AND LEFT MAIN CORONARY ARTERY OCCLUSION

PRIMARNA PCI KOD PACIJENATA SA ANOMALnim ODSTUPOM LEVE KORONARNE ARTERIJE IZ SUPROTNOG VALSALVINOG SINUSA I OKLUZIJOM GLAVNOG STABLA LEVE KORONARNE ARTERIJE

Authors Milenko Čanković*,† Tibor Čanji†, Dragan Debeljački†, Nikola Komazec†, Milovan Petrović*,†, Lazar Velicki*,†, Vojnosanitetski pregled (2021); Online First April, 2021.

UDC:

DOI: https://doi.org/10.2298/VSP201001044C

When the final article is assigned to volumes/issues of the Journal, the Article in Press version will be removed and the final version appear in the associated published volumes/issues of the Journal. The date the article was made available online first will be carried over.
 PRIMARY PCI IN A PATIENT WITH ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE OPPOSITE SINUS OF VALSALVA AND LEFT MAIN CORONARY ARTERY OCCLUSION

PRIMARNA PCI KOD PACIJENTA SA ANOMALNIM ODSTUPOM LEVE KORONARNE ARTERIJE IZ SUPROTNOG VALSALVINOG SINUSA I OKLUZIJOM GLAVNOG STABLA LEVE KORONARNE ARTERIJE

Milenko Čanković*† Tibor Čanji† Dragan Debeljački†, Nikola Komazec†, Milovan Petrović*†, Lazar Velicki*†

*Faculty of Medicine, University of Novi Sad, Novi Sad, Serbia;
†Institute of Cardiovascular Diseases of Vojvodina, Sremska Kamenica, Serbia

Corresponding author:

Asis. Dr. Milenko Čanković

Institute of Cardiovascular Diseases, Vojvodina

Put doktora Goldmana 4

21204 Sremska Kamenica

Tel. +381 21 480 5661

E-mail: milenko.cankovic@mf.uns.ac.rs

Conflict of interest: none declared
Abstract

Introduction. Congenital coronary anomalies are detected in around 5% of all performed coronarographies. Coronary artery anomalies considered as those of great risk are the ones arising from the opposite sinus of Valsalva. These anomalies are detected in around 1% of cases.

This case report shows a unique case of a patient with anterior wall STEMI caused by left main coronary artery (LMCA) occlusion which arises from the right coronary cusp and has interarterial course, which was successfully treated with primary PCI.

Case Report. A 46-year-old male patient was admitted to the hospital due to STEMI of the anterior region. At the admission the patient was hypertensive (150/100 mmHg) in sinus rhythm (heart rate around 70/min), Killip I. After the initial examination and admitting dual antiplatelet therapy, the patient was transferred for urgent coronarography. Coronarography was performed by using the trans-radial approach. The right coronary artery had no significant stenosis and it was easily cannulated, whereas the left coronary artery could not be cannulated at the usual position. The operator attempted to cannulate the left coronary artery with multiple catheters of various curves, unsuccessfully. The conclusion was that there was a coronary artery anomaly, and the cannulation of the anomalous aortic origin of the left coronary artery (AAOLCA) which arises from the opposite right coronary cusp, was successfully performed with a Multipurpose catheter. Moreover, the left coronary artery was occluded in the distal segment. Two drug-eluting stents were implanted and followed by the development of no-reflow phenomenon and cardiogenic shock. After the stabilization, CT coronarography was performed and AAOLCA with an interarterial course was registered. During the follow-up period SPECT was performed and in the staged procedure, the stent was implanted on the proximal Cx, by using the TAP technique.

Conclusion. Patients with STEMI and the anomalies of coronary arteries are very rare. As such, these patients represent a great challenge for revascularization. Possessing the knowledge of anatomic varieties is paramount when it comes to these patients, to treat them adequately with primary percutaneous coronary intervention.

Keywords: anomalous coronary artery; anomalous aortic origin of the left coronary artery; primary percutaneous coronary intervention; ST elevation myocardial infarction.
Sažetak

Uvod. Kongenitalne anomalije koronarnih arterija se sreću do oko 5% svih koronarnografija. Kao posebno rizične smatraju se anomalije ishodišta koronarne arterije iz suprotnog Valsalvinog sinusa (ACAOS) i sreću se u oko 1% slučajeva. Prikazali smo jedinstven slučaj pacijenta sa STEMI prednjeg zida uzrokovanih okluzijom glavnog stabla LCA koja odstupa iz desnog koronarnog kuspisa i ima interarterijski pravac pružanja, a koja je uspešno tretirana primarnom perkutanom koronarnom intervencijom.

Prikaz slučaja. Bolesnik starosti 46 godina primljen je kao hitan slučaj zbog kliničkih i elektrokardiografskih znakova za STEMI anteriorne regije. Po prijemu je hipertenzivan (150/100 mmHg), u sinusom ritmu (FS oko 70/min), Killip I. Nakon inicijalnog pregleda i uvođenja dvojne antiagregacione terapije bolesnik je premešten u kateterizacionu laboratoriju radi urgentne koronarografije. Koronarografija je urađena transradijalnim pristupom, desna koronarna arterija koja je bez signifikantnih suženja je lako kanulisana, dok se leva koronarna arterija ne uspeva kanulisati. Pokušano je sa više katetera različitih krivina, ali bez uspeha. Zaključeno je da se radi o koronarnoj anomaliji i tek sa gajdinig kateterom Multipurpose uspešno je kanulisana leva koronarna arterija koja polazi iz desnog koronarnog kuspisa (AAOLCA), a koja je okludirana u svom distalnom segmentu. Urađena je implantacija dva lekom obložena stenta sa razvojem no reflow-a i kardiogenog šoka. Nakon stabilizacije dva lekom obložena stenta sa razvojem no reflow-a i kardiogenog šoka. Nakon stabilizacije urađena je CT koronarografija kojom je potvrđena AAOLCA, a registrovano je i da ima potencijalno rizični interarterijski pravac kretanja. Nakon urađenog SPECTa tokom perioda praćenja u drugom aktu je urađena implantacija stenta na proksimalnu RCX TAP tehnikom.

Zaključak. Pacijenti sa STEMI i anomalijama koronarnih arterija se relativno retko sreću i predstavljaju izazov za revaskularizaciju, te je poznavanje anatomskih varijeteta neophodno kako bi ovi bolesnici mogli biti adekvatno tretirani sa primarnom perkutanom koronarnom intervencijom.

Ključne reči: anomalna koronarna arterija; anomalno aortno ishodište leve koronarne arterije; primarna perkutana koronarna intervencija, infarkt miokarda sa ST elevacijom.
Introduction
Congenital coronary artery anomalies (CAA) are detected in around 5% of all performed coronarographies [1]. In the literature, there are 66 different anomalies described, while the ones considered of great risk are anomalous coronary arteries from the opposite sinus of Valsalva (ACAOS). These anomalies are detected in around 1% of cases. Anomalous aortic origin of the right coronary artery (AAORCA) arising from the left coronary cusp is detected more often, whereas the anomalous aortic origin of the left coronary artery (AAOLCA) arising from the right coronary cusp is detected in 0.15% of the cases [1-2]. With both CAA, there are several different courses, such as pre-pulmonic, retro-aortic, subpulmonic (septal), or interarterial course where the coronary artery is placed between the aorta and the pulmonary trunk. Interarterial course is the only course that can cause sudden cardiac death and, therefore, is considered malignant [3]. STEMI is rarely detected in patients with coronary artery anomalies and is demanding to treat. The very identification and cannulation of the culprit artery with anomalous origin can be quite challenging.

Herein we presented a unique case of a patient with anterolateral wall STEMI, caused by an occlusion of the left coronary artery arising from the right coronary cusp with interarterial course which was successfully treated with the primary percutaneous coronary intervention (PCI).

Case Report
A 46-year-old patient was admitted to the hospital due to anterior wall STEMI (Figure 1). Chest pain started 2 hours prior to admission to the hospital. The patient had no prior medical history and the only risk factor for coronary artery disease was smoking. At the admission, the patient was hypertensive (150/100 mmHg) in sinus rhythm (HR 70/min), Killip I. After the initial examination, dual antiplatelet therapy was introduced, and the patient was transferred to the Cath lab for urgent coronarography.

Coronarography was performed using trans-radial approach. The right coronary artery (RCA) was easily cannulated with diagnostic catheter Tiger (Terumo, Japan), and it had no significant stenosis, whereas the left coronary artery could not be cannulated in the left coronary cusp. Cannulation of the left coronary artery was
attempted with multiple catheters, such as EBU of various curves, JL 4.0, Amplatz 1, but attempts were unsuccessful.

The operator concluded CAA was in question, and the cannulation of the left coronary artery arising from the right coronary cusp (AAOLCA), which was occluded in its distal segment, was successfully cannulated with a Multipurpose catheter (Mach 1, Boston Scientific, USA). (Figure 2A). After passing the guidewire through the occlusion, the antegrade flow was achieved, followed by ventricular fibrillation and prolonged CPR. Finally, the restoration of spontaneous circulation (ROSC) was established. Due to the progression of heart failure to the state of cardiogenic shock, the patient was intubated and put on mechanical ventilation, and vasopressor support was introduced in the treatment.

Because of the loosening of the position of the guiding catheter and clinical instability, crossover to the right femoral access was performed. This time, LCA was cannulated with a JR 4.0 guiding catheter (Mach 1, Boston Scientific, USA). After passing the guidewire, thrombus aspiration was performed due to the high thrombus burden. Subsequently, predilatation was performed and after that TIMI 2 flow was established. A trifurcation lesion of the left main coronary artery (LMCA) was detected. The plaque was propagating from distal LMCA to the proximal left anterior descending artery (LAD) and circumflex (Cx), while gracile Ramus had no significant lesion at the ostia (Figure 2B). Two drug-eluting stents (DES) were implanted, one DES 3.5 x 23mm (Xience Xpedition, Abbott, USA) from the LMCA to the LAD. The second stent, DES 3.5 x 15mm (Xience Xpedition, Abbott, USA) was implanted in the LMCA, with a short overlap with the previously implanted stent. Furthermore, a proximal optimization technique (POT) with a semi-compliant balloon, 4.0 x 12 mm (Sprinter, Medtronic, USA), with high-pressure inflation was performed. After POT, a no-reflow phenomenon developed (figure 2C), and GP IIb/IIIa inhibitor (Tirofiban) was admitted intracoronary. Also, an intra-aortic balloon pump (IABP) was implanted (Figure 2D), and the patient was transferred to the coronary care unit (CCU). After 48 hours in the CCU, the patient was clinically stable and both vasopressor support and IABP were removed. The patient was extubated after 72 hours. Echocardiography registered an ejection fraction of 40% with akinesia of apex and all apical segments of the left ventricle and hypokinesia of medial anterolateral and inferolateral wall.
On the seventh day of hospitalization, the patient was stabilized and was transferred to the ward. CT coronarography was performed for detailed analyses of the CAA. CT registered anomalous slit-like origin of the left coronary artery from the right coronary cusp (AAOLCA), with the interarterial course (placed between the aorta and pulmonary trunk). Furthermore, the length of the LMCA was 50mm. Interestingly, the artery at the point of 7.2mm from the orifice at the aorta enters the heart muscle and it passes through up to trifurcation, in all its length. The stents were patent and at the Cx ostia significant lesion was registered. (Figure 3). On the fourteenth day of the hospitalization, the patient was discharged in a good general condition. During the six-month follow-up period, the patient had symptoms of stable Angina. SPECT was performed, and it showed significant ischemia in the irrigational area of Cx.

Ten months after the STEMI, recoronorography was performed and this time left Amplatz 1 guiding catheter was used, and successful cannulation of LCA and RCA were achieved. Coronography registered significant stenosis of the ostial Cx, and that the previously implanted stents were patent. As a result, DES 2.5 x 15 mm (Xience Pro, Abbott, USA) was implanted on the ostium of Cx with the TAP technique (Figure 4).

**Discussion**

The case report presented a case of a STEMI patient with anomalous origin of the left coronary artery from the right coronary cusp with interarterial course. Besides, the patient had LMCA occlusion in the distal segment. Best to our knowledge, this is a unique case in the literature.

The usage of noninvasive technologies, such as CT and MRI enable detecting and registering coronary anomalies more frequently [3-4]. The detection of CAA deserves special attention. Even though most of these anomalies is benign, coronary arteries arising from the opposite sinus of Valsalva with interarterial course present a risk of sudden cardiac death. It can be caused by many various factors, such as, slit-like orifice or tangential passage of origin, or, it might result from extreme physical activity which can lead to the compression of the anomalous coronary artery with interarterial course. Extreme physical activity can result in increased blood flow through the aorta and pulmonary artery leading to compression which causes ischemia. It is crucial to emphasize that these anomalies present a great risk for the younger population which is exposed to extreme
physical activity. This mostly refers to sportsmen, athletes, and military recruits. Sometimes, the first manifestation can be sudden cardiac death (SCD), however, very often there are symptoms present during the physical activity, such as angina-like symptoms, arrhythmia, presyncope and syncope [5-6].

The literature provides us with a different frequency of atherosclerotic disease in anomalous coronary arteries when compared to normal. Recent studies indicate a slightly higher incidence of coronary artery disease (CAD) in anomalous coronary arteries than what is shown in earlier researches which indicates an equal incidence of CAD [7]. However, STEMI patients with coronary anomalies are quite rare. According to the research conducted by Marchesini et al, only 5 out of 1015 STEMI patients (0.4%) had an anomaly of the coronary artery at the same time [8].

Performing primary PCI in STEMI patients and CAA is challenging for every operator, mostly because of the cannulation problems of the anomalous culprit artery. However, a problem might arise during the procedure, in terms of balloon and stent deliverability in the culprit lesion area. In this case report, the cannulation of the LCA presented a problem, considering that it could not be detected in the left coronary cusp. After exchanging multiple catheters, the operator decided to use a Multipurpose catheter and search for coronary artery in the right coronary and posterior cusp. Luckily, in this case, only the cannulation was challenging, considering the anomalous origin. In both procedures, there was no problem with the device deliverability.

The occlusion of the left main coronary artery is a disastrous event and most of the patients has a fatal outcome, on the way to the hospital. De Luca et al, registered incidence of 0.8% in patients with a myocardial infarction and the LMCA occlusion. Their study registers high intrahospital mortality, which was 58% in all patients, and 80 % in patients who developed cardiogenic shock or had no re-flow, at the end of the procedure. [9]. Certain factors had shown that they contribute to the higher survival rate of these patients, such as dominant RCA, the existence of hetero-collateral circulation, and fast revascularization [10-12]. In our case, RCA was dominant, and hetero-collateral showed the very periphery of LAD. However, the existence of hetero-collateral circulation did not imply the anomalous origin of LCA. The operator’s experience to assume the presence of coronary anomaly and the adequate selection of catheter allowed successful revascularization of LMCA and proximal LAD.
Despite the loss of time, of around 15 minutes until the cannulation of anomalous LCA, guidewire passage and establishing anterograde flow through the infraction artery, cardiogenic shock developed, as well as, no-reflow at the end of the procedure. No-reflow developed as a consequence of a high thrombus burden and aggressive post-dilatation during POT, which led to distal embolization with thrombus masses and debris from the lesion, causing microvascular obstruction. Treatment of no-reflow was challenging, because nitroglycerin, adenosine or intracoronary adrenalin could not be given due to hemodynamic and rhythmic instability. Decision to administer GP IIb/IIIa (Tirofiban) with implantation of mechanical circulatory support with IABP and aggressive treatment of cardiogenic shock with the support of invasive mechanical ventilation led to a fast stabilization. It is important to emphasize that the VA-ECMO implantation was considered. However, due to the no-reflow and admitting GP IIb/IIIa inhibitor there was a high risk of bleeding, it was decided to wait and monitor the patient’s status. As the satisfactory hemodynamical stabilization was achieved promptly during the treatment at the CCU, the implantation was no longer needed.

Since 3D reconstruction is not possible, angiography has a limited sensitivity when it comes to adequate diagnostics of anomalies of coronary arteries. Therefore, to diagnose the type of anomalies, its course and its placement in relation to great vessels, it is necessary to use MSCT or MRI. Applying these methods prior to coronarography can facilitate the cannulation of the anomalous coronary artery, and this option is manageable in elective and stable patients [13].

Surgical approach to AAOLCA and AAORCA anomalies is considered in the patients younger than 35, and who were exposed to high physical efforts and had experienced some of the symptoms which led to the diagnosis of this type of anomaly. When it comes to older patients, the type of treatment should be carefully considered, especially the need for surgical intervention. Especially, if these patients are asymptomatic. Tests for determining ischemia are advised in these patients [14]. Surgery treatment was not taken into the consideration for the patient presented in this case report since the patient did not have any symptoms before the coronary incident and was not exposed to great physical efforts. SPECT showed ischemia only in the Cx area, which was sub-occluded at the origin and it was revascularized by stent implantation.

**Conclusion**
STEMI patients with coronary artery anomalies are rare and represent a challenge for revascularization, thus, knowing the anatomical varieties is essential to treat these patients with percutaneous coronary intervention.

References

1. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation 2007; 115(10):1296–1305. (English)

2. Angelini P, Flamm SD. Newer concepts for imaging anomalous aortic origin of the coronary arteries in adults. Catheter Cardiovasc Interv 2007; 69(7): 942-954. (English)

3. Perez-Pomares JM, de la Pompa JL, Franco D, Henderson D, Ho SY, Houyel L, et al. Congenital coronary artery anomalies: a bridge from embryology to anatomy and pathophysiology—a position statement of the development, anatomy, and pathology ESC Working Group. Cardiovasc Res 2016; 109(2): 204-216. (English)

4. Garg N, Tewari S, Kapoor A, Gupta DK, Sinha N. Primary congenital anomalies of the coronary arteries: a coronary arteriographic study. International Journal of Cardiology 2000; 74(1): 39–46. (English)

5. Eckart RE, Scoville SL, Campbell CL, Shry EA, Stajduhar KC, Potter RN, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med 2004; 141(11): 829-834. (English)

6. Roberts WC. Major anomalies of coronary arterial origin seen in adulthood. Am Heart J 1986; 111(5): 941-963. (English)

7. Sidhu NS, Wander GS, Monga A, Kaur A. Incidence, Characteristics and Atherosclerotic Involvement of Coronary Artery Anomalies in Adult Population Undergoing Catheter Coronary Angiography. Cardiol Res 2019; 10(6): 358–368. (English)

8. Marchesini J, Campo G, Righi R, Benea G, Ferrari R. Coronary artery anomalies presenting with ST-segment elevation myocardial infarction. Clin Pract 2011 28; 1(4): e107. (English)

9. De Luca G, Suryapranata H, Thomas K, van ’t Hof AW, de Boer MJ, Hoortje JC et al. Outcome in patients treated with primary angioplasty for acute myocardial infarction due to left main coronary artery occlusion. Am J Cardiol 91(2): 235-238. (English)
10. Shigemitsu O, Hadama T, Miyamoto S, Anai H, Sako H, Iwata E. Acute myocardial infarction due to left main coronary artery occlusion. Therapeutic strategy. Jpn J Thorac Cardiovasc Surg 2002; 50(4): 146–151. (English)

11. Spiecker M, Erbel R, Rupprecht HJ, Meyer J. Emergency angioplasty of totally occluded left main coronary artery in acute myocardial infarction and unstable angina pectoris--institutional experience and literature review. Eur Heart J 1994; 15(5): 602–607. (English)

12. Li JJ, Xu B, Chen JL. Stenting for left main coronary artery occlusion in adolescent: A case report. World J Cardiol 2010; 2(7): 211–214. (English)

13. Ten Kate GJR, Weustink AC, de Feyter PJ. Coronary artery anomalies detected by MSCT-coronary angiography in the adult. Neth Heart J 2008; 16(11): 369–375. (English)

14. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA et al. A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults with Congenital Heart Disease): Developed in Collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. Circulation 2008; 118(23): 2395–2451. (English)
**Figure 1.** ECG at admission.

**Figure 2** A) The arrow points to the occlusion site of the distal segment AAOLCA. LAO 46, CAU 3; B) Anterograde flow is established, and a trifurcation lesion is detected. RAO 2, CRA 26. C) No-reflow at the end of the procedure. RAO 14, CRA 26. D) Positioning of IABP. AP 0
Figure 3  A) The arrow points to the anomalous origin of LCA from the opposite sinus of Valsalva. B) 3D reconstruction registers the interarterial course of AAOLCA, arrow point to the part of the artery which is inside of the muscle. (Ao–aorta, PA–pulmonary artery). C) The left arrow points to the slit-like orifice of AAOLCA, the right arrow points to significant stenosis of ostial Cx.

Figure 4  A) The arrow points to a lesion at the Cx ostia RAO 46 CRA 30. B) TAP technique, DES 2.5 x 15 mm in Cx and NC balloon.5 x 12 mm in LMCA -LAD3RAO 46, CRA 30. C) Final result LAO 7, CRA 25.
Received on October 1, 2020.
Revised on December 13, 2020.
Accepted April 20, 2021.
Online First April, 2021.