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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Anomalous aortic origin of right and circumflex coronary arteries – procedural risks during combined aortic valve replacement and coronary artery bypass grafting

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SUMMARY

Introduction Anomalous aortic origin of the right and the circumflex coronary arteries presents extremely rare and potentially dangerous combination in patients scheduled for combined coronary bypass grafting and aortic valve replacement surgery. We report this illustrative case to emphasize the importance of meticulous diagnostic setup enabling the surgeon to anticipate and avoid numerous possible pitfalls.

Case outline A 74-year-old woman, with anterior-wall myocardial infarction and aortic valve stenosis, underwent successful combined coronary artery bypass grafting and aortic valve replacement. Preoperative coronary angiography revealed unusually high take-off of the right main coronary trunk and anomalous origin and course of the circumflex coronary artery. Anatomy of both anomalous coronary arteries in the light of underlying surgical pathology necessitated a meticulous preparation and caution during successive phases of surgical treatment.

Conclusion Estimating potential procedural risk should be standard practice for each patient with known congenital coronary artery anomalies, regardless of the natural risk imposed by a particular anomaly. Preoperative evaluation of coronary circulation, with high surgical awareness and knowledge of different congenital coronary artery anomalies, should be a standard approach in cardiac surgical practice. This would add a predictive value for an actual procedural risk in cases of previously unrecognized anomalies.

Keywords: coronary vessel anomalies; cardiac surgical procedures; heart valve prosthesis implantation; myocardial revascularization

INTRODUCTION

The precise risk stratification for patients with isolated (primary) congenital coronary artery anomalies (CCAA) is difficult to determine, and thus management decisions should be highly individualized [1, 2]. For practical reasons, we propose two mayor risk categories to be considered and individually estimated:

1) Natural risk – defining whether and how particular CCAA, *per se*, imposes increased morbidity and/or mortality risk to the patient;
2) Procedural risk – anticipating possible surgical and/or interventional pitfalls and hazards related to the presence of CCAA.

These two risk categories do not inevitably coincide. Thus, CCAA with low or nonexistent natural risk may bring significant procedural risk for the patients scheduled for certain cardiac surgical and/or interventional procedures.

Anomalous aortic origin of the right (RCA) and the circumflex (aCx) coronary arteries – the former with high aortic take-off and the latter branching from the proximal RCA, adopting anomalous retroaortic course – present an extremely rare and potentially a very dangerous combination in adult patients scheduled for combined coronary bypass grafting and aortic valve replacement (AVR) [3, 4, 5]. Although the natural risk of such CCAA combination is commonly reported to be low, particular procedural risk is significant. We report this illustrative case to emphasize the importance of meticulous diagnostic setup and interpretation, enabling the surgeon to anticipate and avoid numerous possible pitfalls.

CASE REPORT

A 74-year-old woman, with a history of aortic stenosis, hypertension, and diabetes, was admitted with an acute, anterior-wall, non-ST elevating myocardial infarction. Transthoracic echocardiography revealed the tricuspid, severely calcified, stenotic aortic valve, with an orifice area of 0.74 cm^2 and mean/maximal systolic pressure gradient of 83/116 mmHg. Left-side coronary angiography showed the left anterior descending coronary artery (LAD), with proximal 50% area stenosis and "absent Cx." Right-side coronary angiography did not depict RCA at the usual position, but it emerged as a long common RCA trunk (RCT), having wide, slit-like orifice, 1.5 cm above the sinotubular junction (STJ), giving a rise to small and retroaortic aCx (Figure 1). The definitive preoperative

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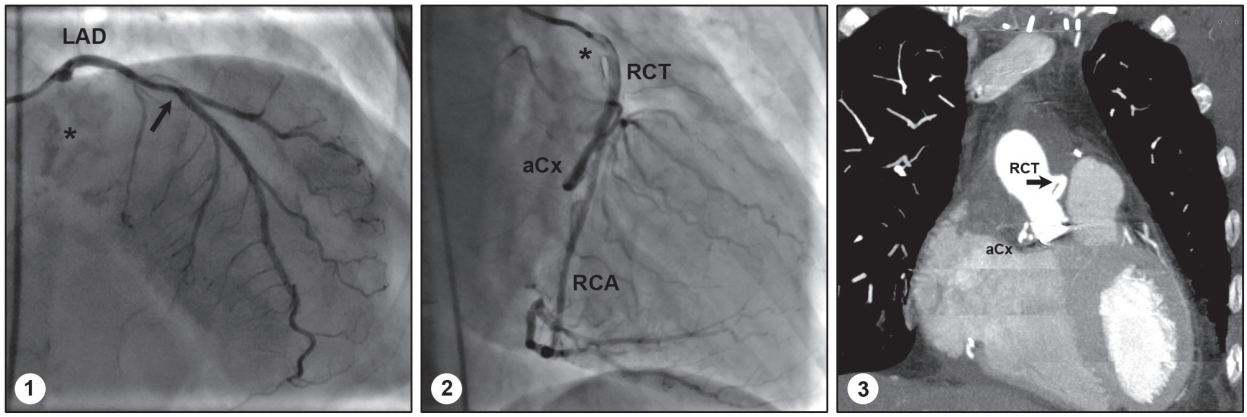

Figure 1. Coronary contrast imaging: 1) a separate left anterior descending coronary artery origin from the left sinus of Valsalva, with 50% proximal stenosis (arrow); stenosed and calcified aortic valve (asterisk); 2) long right coronary artery trunk (RCT) with high ascending aortic take-off and wide slit orifice (asterisk); (3) multislice computerized tomography scan depicting the RCT ascending aortic orifice (arrow) and course

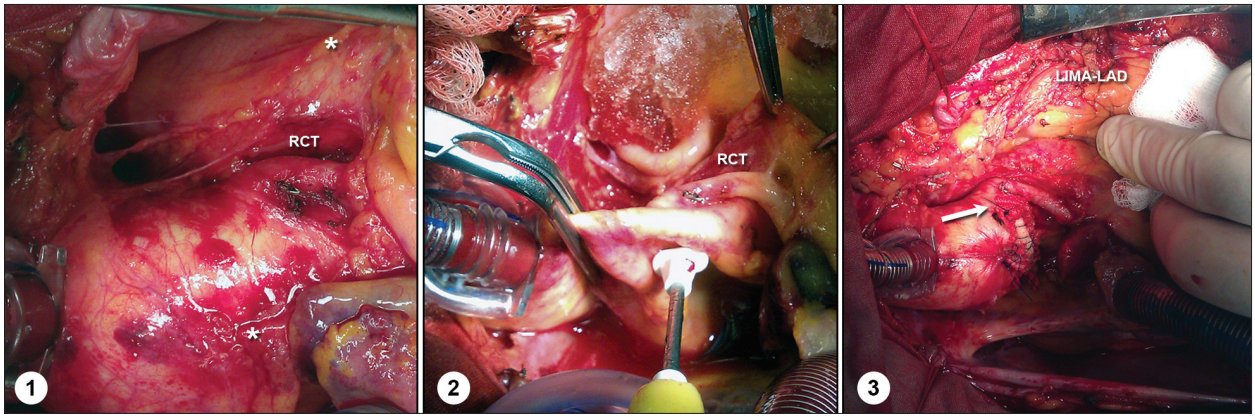

Figure 2. Intraoperative (surgeon's view): 1) ascending aortic ridge (asterisks) dissected to expose the right coronary artery trunk (RCT) high take-off; 2) narrow surgical field between the aortic cross-clamp and the RCT; 3) modified small-sized aortotomy leaving a free wall space (arrow) toward the RCT orifice

evaluation was supplemented with stress-ECHO and coronary flow reserve (CFR) assessment, revealing reduced anterior wall motion and CFR = 1.7 on the distal LAD. The patient was scheduled for combined CABG-LAD and AVR.

Upon standard median sternotomy and heart exposure, no apparent evidence of any congenital heart or CCAA was present. The ascending aorta, above the Rindfleisch's ridge, appeared short and narrow. Palpation and careful dissection of the ascending aortic ridge, toward the aorto-pulmonary groove, exposed a long, extramural, common RCT, originating 1.5 cm above the STJ, leaving a very narrow portion of the ascending aorta free for arterial cannulation, aortic cross-clamping, cardioplegic needle-cannula insertion, and aortotomy. The left internal mammary artery (LIMA) for LAD grafting was guided not only on the basis of its well established preferences, but also by anticipated spatial limitations for the eventual proximal anastomosis. Modified, small, right-sided aortotomy was created, to ensure a feasible aortic valve exposure, leaving sufficient space around the RCT orifice (above the commissure between the left and right aortic cusp), for the subsequent safe aortotomy closure. Special attention was paid to repeated cardioplegic infusions, in order to adjust direct cannula tip size and orientation, with wide, slit-like RCT orifice and its tangential course, over the bulbar aortic portion. Valvular decalcification and a 19 mm St. Jude Medical™ Regent™ mechanical aortic prosthesis implantation (St. Jude Medical Inc., St. Paul, MN, USA) was uncomfortable and challenging, not only because of limited space, but also because of retroaortic course of the aCx and unusually low LAD orifice (Figure 2). The entire procedure was uneventful and the patient was easily weaned from cardiopulmonary bypass.

Soon after initial recovery, a multislice computerized tomography (MSCT) scanning was performed. It confirmed, in a more picturesque and precise manner, all our concerns and anticipations, provided by preoperative coronary angiography and intraoperative exploration (Figure 3). Moreover, it also confirmed the absence of retroaortic aCx compression by the prosthetic sewing ring, as well as good patency of other native coronary arteries and LIMA-LAD graft. The patient was discharged on the seventh postoperative day and is still doing well, without any angina pectoris and/or dyspnea, during the 4.5-year-long follow-up.

DISCUSSION

Appreciation of the "procedural risk" (as defined here), related to CCAA, has evolved little since the earliest

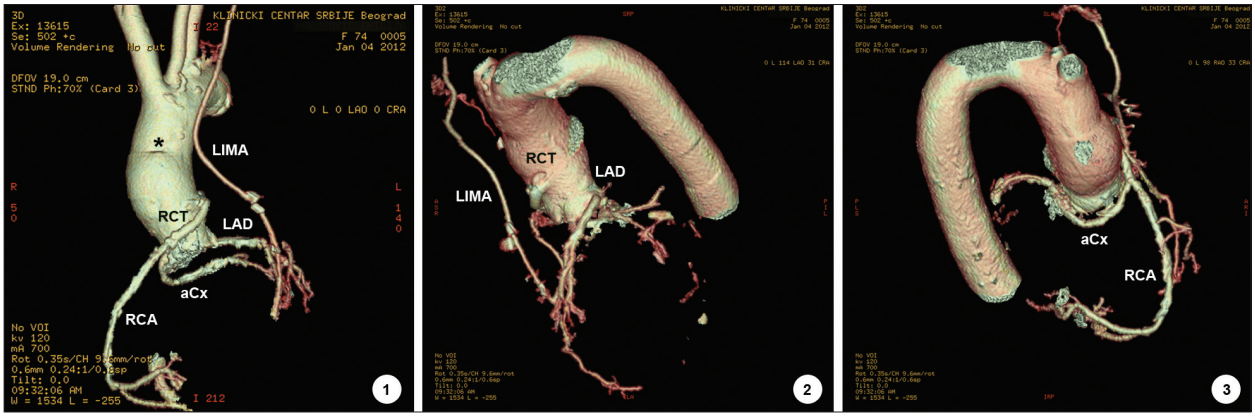

Figure 3. Postoperative 64-slice computed tomography scan: 1) anterior view, depicting the right coronary artery trunk (RCT) origin, course and branching. Asterisk indicates aortic cannulation site; 2) left anterior oblique view, showing a spatial relation between the RCT and left anterior descending coronary artery orifices; 3) left posterior oblique view, depicting anomalous origin and retroaortic course of aCx

observations by Vlodaver et al. [6], and later on by Blake et al. [7] and Angelini [8]. On the other hand, "natural risk" associated with CCAA has usually been of main concern in numerous classifications during the last three decades. As a result, the clinical attention to isolated (primary) CCAA with low or no natural risk has slacked within cardiac surgical community [4, 5, 8, 9, 10].

Angelini et al. [1, 11] defined the minimum criteria describing normal coronary artery network, suggesting that term "anomalous" should reflect only the anatomy, which is present in less than 1% of the general population. In the population undergoing coronary angiography, the prevalence of CCAA is reported to be 1.3%, while in unselected autopsy series they are present in 0.3% of cases [1, 11]. Noteworthy, in autopsies of young athletes, CCAA were found in 11.8–19% of cases, being the second most common cause of sudden cardiac death in this population [12, 13, 14]. The true prevalence of CCAA in the general population is difficult to establish but it is probably much higher than reported, as the majority of them impose no limitations on resting or maximal blood flow and thus remain clinically silent.

Since the earliest attempt to classify different CCAA, integrating anatomical, functional and clinical features, by Vlodaver et al. [6] and Edwards [15], different classifications have appeared (e.g. Ogden, Angelini), and still none have been widely accepted [2, 16–21]. Critical attitude in interpreting "casual vs. causal" relationship in CCAA is of utmost importance [8, 19, 20, 21].

Among all variations of the position of coronary artery orifices (in the frontal plane), a high RCA and a low left coronary artery orifice is the second rarest combination, encountered in 8% of the cases. Loukas et al. [22] proposed that "high take-off" denomination should refer only to anomalous aortic origin of the coronaries (AAOC) arising ≥ 1 cm above the STJ, as in our case. Applying strictly this criterion in their meta-analysis, they reported the prevalence of 0.20% (26 of 12,899 cases), with RCA being the most common (84.46%) high take-off artery. Sudden cardiac death was recorded in 0.02% of the cases. Acute angle high take-off, together with interarterial and/or intramural course may alter coronary blood flow, even in the absence of atherosclerosis [22]. The first two of these three characteristics, as noted above, were also present in our patient.

The AAOC with aCx branching from the RCA (RCT) was first reported by Antopol and Kugel in 1933 [5]. This is one of the most common CCAA, with prevalence reported in angiographic series ranging from 0.45% to 0.70%. The first case of aCx in our national pathology was described in 1964 by Kanjuh and associates [23]. In a recent study of myocardial bridges, Teofilovski-Parapid et al. [24] have found this anomaly present in 7.7% of the 96 hearts studied. The aCx in our patient arose as a discrete proximal branch of a long anomalous RCT, which is the most common of three possible aCx branching patterns [5].

Taking into account the advanced age and available preoperative data, we could not find any firm functional correlation between described anatomical features of the RCT (RCA) and aCx with the past or presenting clinical presentation. Whether particular CCAA combination in the absence of obstructive atherosclerosis carries low or no natural risk remains difficult to document in the absence of previous functional testing.

Yet, procedural risk associated with scheduled combined cardiac surgical procedure for this particular patient was very high. Not many papers have described interventional or surgical challenges for either RCA high aortic take-off or retroaortic aCx branching from the RCA, and we found even less reports describing the presence of these CCAA in a setting of combined AVR and CABG [25–31].

Potential traps during the coronary angiography of such patients include "missing Cx" and difficult access to the RCA orifice. Whenever there are no traces of Cx on standard left angiograms (a sign of nonperfused myocardium), one should think about the possibility of aCx presence. Also, inability to find and access the RCA orifice within the right Valsalva sinus, using standard catheters, should raise suspicion of high aortic RCA take-off [25, 26].

In available and very rare surgical reports, AAOC, including isolated or combined high aortic RCA take-off (according to Loukas' crteria) and retroaortic aCx branching from the RCA was often recognized as CCAA with

significant procedural risk, depending on the cardiac surgical procedure. The most common procedural risks, reported for high aortic RCA take-off, were its damage during inadvertent preparation of the aortopulmonary groove, occlusion by aortic cross-clamp, or transection during the aortotomy for AVR [3, 4, 9, 27–30]. Retroaortic aCx, branching from the RCA, has been reported as the procedural risk mainly during the aortic and/or mitral valve surgery, due to injury caused by the valvular suture placement or to compression by the prosthetic valve ring [5, 31]. Difficulties related to myocardial protection in patients with these CCAA have not been reported so far, but we have stressed some important steps in ensuring adequate and safe cardioplegia delivery.

Estimating potential procedural risk should be standard practice for each patient with known CCAA, regardless of natural risk imposed by particular anomaly. Also, meticulous evaluation of coronary circulation, with high surgical awareness and knowledge of different CCAA, should be a standard approach in preoperative cardiac surgical practice. This would add a predictive value for the actual procedural risk in cases of previously unrecognized CCAA It is time, in our opinion, to reconsider contemporary practice of submitting younger patients to routine valvular or congenital cardiac operations without preoperative insight into coronary artery anatomy (by either angiography or MSCT).

REFERENCES

1. Angelini P. Coronary artery anomalies – current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guidelines. Tex Heart Inst J. 2002; 29(4):271–8.
2. Rigatelli G, Docali G, Rossi P, Bandello A, Rigatelli G. Validation of a clinical-significance-based classification of coronary artery anomalies. Angiology. 2005; 56(1):25–34.
3. Abdulbaki AM, Shah S. Origin of right coronary artery from the ascending aorta: an extremely rare anomaly. Int J Clin Cardiol. 2015; 2:041.
4. Tsai WL, Wei HJ, Tsai IC. High-take-off coronary artery: a haemodynamically minor, but surgically important coronary anomaly. Pediatr Radiol. 2010; 40(2):232–3.
5. Page HL Jr, Engel HJ, Campbell WB, Thomas CS Jr. Anomalous origin of the left circumflex coronary artery. Recognition, antiographic demonstration and clinical significance. Circulation. 1974; 50(4):768–73.
6. Vlodaver Z, Neufeld HN, Edwards JE. Pathology of coronary disease. Semin Roentgenol. 1972; 7(4):376–94.
7. Blake HA, Manion WC, Mattingly TW, Baroldi G. Coronary Artery Anomalies. Circulation. 1964; 30:927–40.
8. Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation. 2007; 115(10):1296–305.
9. Singh D, Darbari A, Sharma MK, Parikh N. Anomalous high origin of right coronary artery above the sinotubular junction: rarely diagnosed anomaly. BMJ Case Rep. 2010; 2010.
10. Chiu IS, Anderson RH. Can we better understand the known variations in coronary arterial anatomy? Ann Thorac Surg. 2012; 94(5):1751–60.
11. Angelini P, Villason S, Chan AV, Diez JG. Normal and anomalous coronary arteries in humans. In: Angelini P, Fairchild VD, editors. Coronary artery anomalies: a comprehensive approach: Lippincott Williams & Wilkins; 1999. p. 27–79.
12. Bille K, Figueiras D, Schamasch P, Kappenberger L, Brenner JI, Meijboom FJ, et al. Sudden cardiac death in athletes: the Lausanne Recommendations. Eur J Cardiovasc Prev Rehabil. 2006; 13(6):859–75.
13. Brothers JA, Paridon SM. The new AHA/ACC guidelines for competitive sports participation in young athletes with anomalous coronary arteries: the evolution of change. World J Pediatr Congenit Heart Surg. 2016; 7(2):241–4.
14. Higgins JP, Andino A. Soccer and Sudden cardiac death in young competitive athletes: a review. J Sports Med. 2013; 2013:967183.
15. Edwards JE. Anomalous coronary arteries with special reference to arteriovenous-like communications. Circulation. 1958; 17(6):1001–6.
16. Gross L. Variation in distribution of the coronary arteries. In: Gross L, editor. The blood supply to the heart in its anatomical and clinical aspects. New York: P.B. Hoeber; 1921. p. 26–37.
17. Yuan SM. Anomalous origin of coronary artery: taxonomy and clinical implication. Rev Bras Cir Cardiovasc. 2014; 29(4):622–9.
18. Jacobs ML, Mavroudis C. Anomalies of the coronary arteries: nomenclature and classification. Cardiol Young. 2010; 20 Suppl 3:15–9.
19. Angelini P. A casual versus causal relationship in coronary artery anomalies: a question of method. Tex Heart Inst J. 2004; 31(3):276–7.
20. Brothers JA, Gaynor JW, Jacobs JP, Caldarone C, Jegatheeswaran A, Jacobs ML. The registry of anomalous aortic origin of the coronary artery of the Congenital Heart Surgeons' Society. Cardiol Young. 2010; 20 Suppl 3:50–8.
21. Poynter JA, Williams WG, McIntyre S, Brothers JA, Jacobs ML. Anomalous aortic origin of a coronary artery: a report from the Congenital Heart Surgeons Society Registry. World J Pediatr Congenit Heart Surg. 2014; 5(1):22–30.
22. Loukas M, Andall RG, Khan AZ, Patel K, Muresian H, Spicer DE, et al. The clinical anatomy of high take-off coronary arteries. Clin Anat. 2016; 29(3):408–19.
23. Velimirovic D, Kanjuh V, Tucakovic G, Lastic-Maletic S, Vasiljevic JD. Pathologic morphology and clinical significance of the anomalous origin of the left circumflex coronary artery from the right coronary artery. General review and autopsy analysis of 30 cases. Glas Srp Akad Nauka Med. 1990(39):77–103.
24. Teofilovski-Parapid G, Jankovic R, Kanjuh V, Virmani R, Danchin N, Prates N, et al. Myocardial bridges, neither rare nor isolated – autopsy study. Ann Anat. 2017; 210:25–31.
25. Prakash M, Puri SK, Sethi S, Verma AK, Yadav R, Gupta S, et al. Aberrant and anomalous coronary arteries: What a watchful radiologist's eye shouldn't miss! ERC 2015; Vienna, Austria, 2015.
26. Neves PO, Andrade J, Moncao H. Coronary anomalies: what the radiologist should know. Radiol Bras. 2015; 48(4):233–41.
27. Goena C, Ibarra A, Campana M, Natividad R, Garcia K, Montes PM. High risk anomalous origin of the right coronary artery. Rev Esp Cardiol (Engl Ed). 2012; 65(4):383–4.
28. Nishi H, Mitsuno M, Tanaka H, Ryomoto M, Fukui S, Miyamoto Y. High anomalous origin of the right coronary artery associated with aortic stenosis: a word of caution. Ann Thorac Surg. 2010; 89(3):961–3.
29. Motamedi MH, Hemmat A, Kalani P, Rezaee MR, Safarnezhad S. High take-off of right coronary artery: an extremely rare case of RCA anomaly. J Card Surg. 2009; 24(3):343–5.
30. Gaudino M, Glieca F, Bruno P, Piancone FL, Alessandrini F, Possati G. Unusual right coronary artery anomaly with major implication during cardiac operations. Ann Thorac Surg. 1997; 64(3):838–9.
31. Vaishnava P, Pyo R, Filsoufi F, Sharma S. Compression of an anomalous left circumflex artery after aortic and mitral valve replacement. Ann Thorac Surg. 2011; 92(5):1887–9.

Аномално аортно порекло десне и циркумфлексне коронарне артерије – процедурални ризици током комбиноване замене аортног залиска и коронарног бајпаса

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САЖЕТАК

Увод Аномално аортно порекло десне и циркумфлексне коронарне артерије представља екстремно ретку и потенцијално опасну комбинацију код болесника предвиђених за комбиновану операцију реваскуларизације миокарда и замене аортног залиска.

Циљ нам је да приказом овог илустративног случаја нагласимо важност пажљиве дијагностичке обраде, која омогућава хирургу да предвиди и избегне бројне замке током операције.

Приказ болесника Жена стара 74 године, са акутним инфарктом предњег зида и аортном стенозом, подвргнута је успешној комбинованој операцији реваскуларизације миокарда и замене аортног залиска. Преоперативна коронарна ангиографија је указивала на постојање необично високог аортног одступа стабла десне и аномалног порекла и тока циркумфлексне коронарне артерије. Анатомија обе аномалне коронарне артерије, у светлу постојеће кардиохируршке патологије, захтевала је прецизну припрему и опрезност у свим фазама хируршког лечења.

Закључак Процена могућег процедуралног ризика би требало да буде стандардна пракса за све кардиохируршке болеснике са познатим конгениталним аномалијама коронарних артерија, без обзира на природни ризик који свака од њих носи. Преоперативна евалуација коронарне циркулације, уз висок степен позорности и познавања различитих конгенитаних аномалија коронарних артерија, требало би да буде стандардни приступ у кардиохируршкој пракси. Тиме би се повећала могућност предвиђања реалног процедуралног ризика и код болесника са претходно недијагностикованим аномалијама.

Кључне речи: аномалије коронарних артерија; кардиохируршке процедуре; уградња вештачких срчаних залистака; реваскуларизација миокарда