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Address: 1 Kraljice Natalije Street, Belgrade 11000, Serbia
☏ +381 11 4092 776, Fax: +381 11 3348 653
E-mail: office@srpskiarhiv.rs, Web address: www.srpskiarhiv.rs

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Case Report / Приказ болесника

Mladen Kočica[1,2†], Milica Karadžić[1,2], Miloš Grujić[1,2], Dragan Cvetković[1,2], Ljiljana Šoškić[1,2]

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

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

[1]Clinical Centre of Serbia, Clinic for Cardiac Surgery, Belgrade, Serbia;
[2]University of Belgrade, Faculty of Medicine, Belgrade, Serbia

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† **Correspondence to:**
Mladen KOČICA
Clinical Centre of Serbia, Clinic for Cardiac Surgery, 8 Kosta Todorovic St., Belgrade 11000, Serbia
Email: **kocica@sbb.rs**

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

SUMMARY

Introduction Anomalous aortic origin of the right and the circumflex coronary arteries present 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 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 a standard practice for each patient with known congenital coronary artery anomalies, regardless of natural risk imposed by 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 actual procedural risk in cases of previously unrecognized anomalies.

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

САЖЕТАК

Увод Аномална аортна порекла десне и циркумфлексне коронарне артерије представљају екстремно ретку и потенцијално опасну комбинацију код пацијената, 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 Особа женског пола, стара 74 године, са акутним инфарктом предњег зида и аортном стенозом, подвргнута је успешној комбинованој операцији реавскуларизације миокарда и замене аортног залиска. Преоперативна коронарна ангиографија је указивала на постојање необично високог аортног одступа стабле 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 потенцијалног процедуралног ризика би требало да буде стандардна пракса за све кардиохируршке пацијенте са познатим конгениталним аномалијама коронарних артерија, без обзира 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 пацијената са претходно недијагностикованим аномалијама.

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

INTRODUCTION

The precise risk stratification for the patients with isolated (primary) congenital coronary artery anomalies (CCAA) is difficult to determine, and thus management decisions should be highly

individualized [1, 2]. For the practical reasons, we propose two mayor risk categories to be considered and individually estimated:

1) Natural risk - defining whether and how does particular CCAA, *per se*, impose 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 none natural risk, may bring significant procedural risk for the patients scheduled for certain cardiac surgical and/of 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 extremely rare and potentially very dangerous combination in adult patient, scheduled for combined coronary bypass grafting (CABG) and aortic valve replacement (AVR) [3, 4, 5]. Although 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 surgeon to anticipate and avoid numerous possible pitfalls.

CASE REPORT

A 74-year-old woman, with history of aortic stenosis, hypertension and diabetes, was admitted with an acute, anterior-wall, non-ST elevating myocardial infarction. Transthoracic echocardiography (TT-ECHO) revealed 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 left anterior descending coronary artery (LAD), with proximal 50% area stenosis and "absent Cx". Right side coronary angiography did not depict RCA at usual position, but it emerged as long common RCA trunk (RCT), having wide, slit-like orifice, 1.5cm above sinotubular junction (STJ), giving a rise to small and retroaortic aCx (Figure 1). Definitive preoperative evaluation was supplemented with stress-ECHO and coronary flow reserve (CFR) assessment, revealing reduced anterior wall motion and CFR=1.7 on 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. Ascending aorta, above the Rindfleisch's ridge, appeared short and

narrow. Palpation and careful dissection of the ascending aortic ridge, towards aortopulmonary groove, exposed a long, extramural, common RCT, originating 1.5cm 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. 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 on 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 mechanical, 19mm, St. Jude Medical™ Regent™ (St. Jude Medical Inc. St. Paul, MN, USA) aortic prosthesis implantation, 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). Entire procedure has gone uneventfully and 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 seventh postoperative day and is still doing well, without any angina pectoris and/or dispnea, during the 4.5 year long follow-up.

DISCUSSION

Appreciation of „procedural risk" (as defined here), related to CCAA, has evolved a little since the earliest observations by Jessie Edwards, and later on by Blake and associates [6, 7, 8]. On the other hand, "natural risk" associated with CCAA has usually been of main concern in numerous classifications during the last 3 decades. As a result, the clinical attention to isolated (primary) CCAA with low or none natural risk has slacked within cardiac surgical community [4, 5, 8, 9, 10].

Angelini and associates, 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 general population. In 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 general population is difficult to establish but it's probably much higher than reported, because 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 Edwards and associates [6, 15], different classifications have appeared (e.g. Ogden, Angelini) and still, none have been widely accepted [2, 16–21]. Critical attitude in interpreting "casual *versus* causal" relationship in CCAA is of utmost importance [8, 19, 20, 21].

Among all variations on the position of coronary artery orifices (in the frontal plane), a high RCA and a low LCA orifice is the second rarest combination, encountered in 8% of cases. Loukas and associates proposed that "high take-off" denomination should refer only to AAOC arising ≥ 1 cm above 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% 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 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 and associates have found this anomaly present in 7.7% cases, among 96 hearts studied [24]. The aCx in our patient arose as a discrete proximal branch of long anomalous RCT, which is the most common of 3 possible aCx branching patterns [5].

Taking into account an advanced age and available preoperative data, we could not find any firm functional correlation between described anatomical features of RCT (RCA) and aCx with past or presenting clinical presentation. Whether particular CCAA combination, in the absence of obstructive atherosclerosis, carries out low or none 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 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 of RCA orifice. Whenever there are no traces of Cx on standard left angiograms (sign of nonperfused myocardium), one should think about possibility of aCx presence. Also, inability to find and access RCA orifice within right Valsava 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 RCA was often recognized as CCAA with significant procedural risk, depending on cardiac surgical procedure. The most common procedural risks, reported for high aortic RCA take-off were: its damage during inadvertent preparation of aortopulmonary groove; occlusion by aortic cross-clamp; or transection during the aortotomy for AVR [3, 4, 9, 27–30]. Retroaortic aCx, branching from RCA, has been reported as procedural risk mainly during the aortic and/or mitral valve surgery, due to: injury caused by the valvular suture placement; or, compression by 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 out some important steps in ensuring adequate and safe cardioplegia delivery.

Estimating potential procedural risk should be a 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 actual procedural risk in cases of previously unrecognized CCAA It is a time, in our opinion, to reconsider contemporary practice of submitting younger patients to routine valvular or congenital cardiac operations without preoperative insight in coronary artery anatomy (by either angiography or MSCT).

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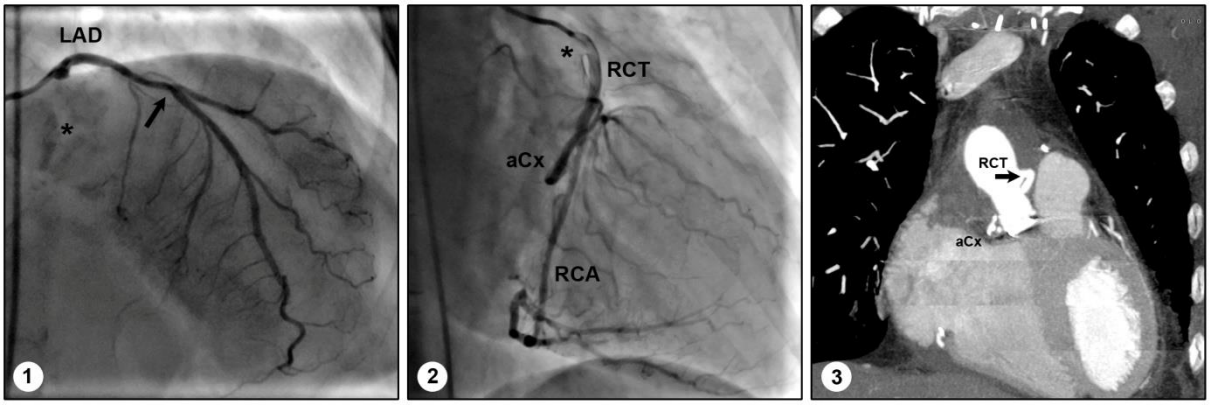

Figure 1. Coronary contrast imaging: (1) – A separate LAD origin from the left sinus of Valsalva, with 50% proximal stenosis (arrow). Stenosed and calcified aortic valve (asterisk). (2) – Long RCT with high ascending aortic take-off and wide slit orifice (asterisk). (3) – MSCT scan depicting RCT ascending aortic orifice (arrow) and course

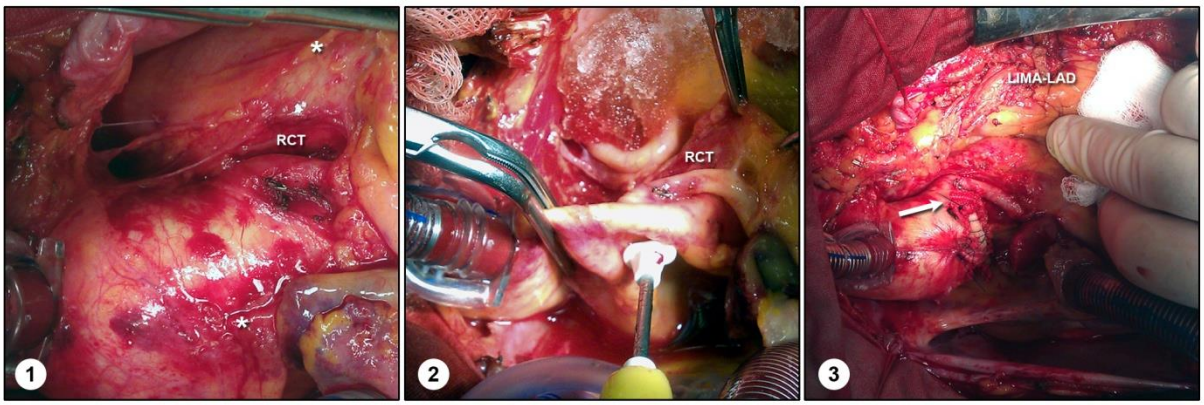

Figure 2. Intraoperative (surgeon's view): (1) - Ascending aortic ridge (asterisks) dissected to expose the RCT high take-off. (2) – Narrow surgical field between aortic cross-clamp and RCT. (3) – Modified small-sized aortotomy leaving a free wall space (arrow) towards the RCT orifice

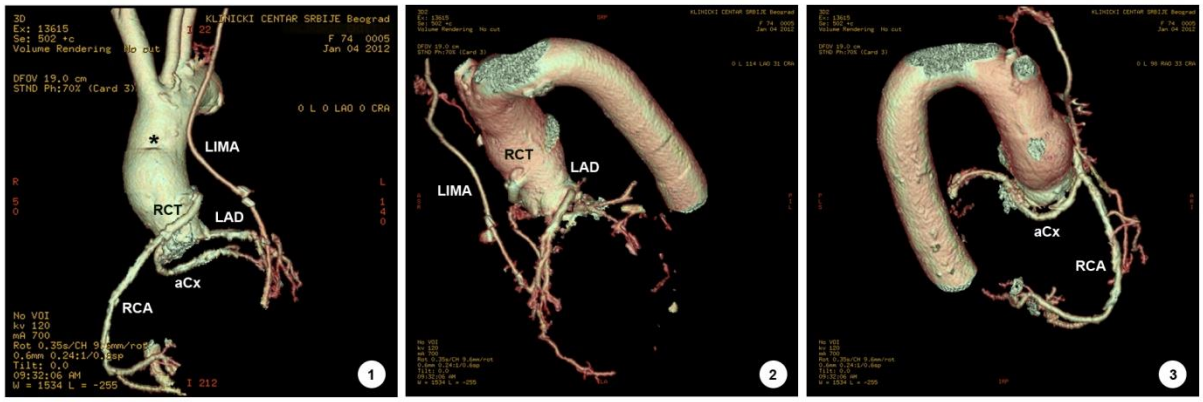

Figure 3. Postoperative 64-slice computed tomography scan: (1) – Anterior view, depicting RCT origin, course and branching. Asterisk indicates aortic cannulation site. (2) Left anterior oblique view, showing a spatial relation between RCT and LAD orifices. (3) Left posterior oblique view, depicting anomalous origin and retroaortic course of aCx

