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Case Report

Multiple Coronary Artery Bypass Grafting in a Patient with Dextrocardia and Situs Viscerum Inversus Totalis

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Abstract

Dextrocardia with situs inversus is a rare congenital anomaly in which the heart and the abdominal organs orient themselves in a mirror-image reversal of the normal anatomy. Coronary artery disease incidence is similar to that of the normal population. Performing coronary artery bypass grafting in this subset of the population poses few difficulties. These limitations can be overcome by few technical adjustments by the surgeon and the team which will be discussed in our article. Coronary artery bypass surgery (CABG) in dextrocardia with situs inversus patients is reported less in literature. Due to abnormal looping and associated other congenital anomalies, anesthetic implications and surgical difficulties become challenging in these patients. The article presents a successful case of multiple coronary artery bypass grafting in a patient with dextrocardia and situs inversus totalis.

Keywords: Dextrocardia; Situs Inversus; Coronary Artery Bypass Grafting

Dextrocardia is an intrauterine anomaly when the major part of the heart is located in the right part of the thorax. Dextrocardia belongs to rare anomalies: the incidence rate varies in different regions from 1:8000 to 1:25000 newborns. In Russia, the prevalence of dextrocardia is 3.3% of all anomalies concerning the position of the organ.

Situs viscerum inversus is a rare abnormal development resulting from disorders of fetal egg differentiation and manifesting itself in mirrored, in relation to the normal, arrangement of internal organs. The incidence rate varies from 1:5000 to 1:20000. Such position of inner organs is observed in one case per 10 million births [1]. The specific genetic cause of dextrocardia with situs inversus was not identified, so patterns of inheritance were not confirmed in the majority of cases, but the condition results from disorders of the implementation of left-right-sided asymmetry in the prenatal period [1].

Dextrocardia combined with situs viscerum inversus is a rare congenital anomaly with the incidence 1:10000 [2,3]. Patients with SIT rarely have cardiac disorders, meanwhile, disorders related to isolated dextrocardia are observed more often. The incidence of coronary artery disease in these patients is relevant to the incidence across the population. Coronary artery bypass grafting in this group of patients is complicated by some difficulties and peculiarities. Hieronymus Facius, an Italian anatomist and surgeon, was the first to describe heart misplacement in the thorax in 1606. Marco Aurelio Severino used the term "dextrocardia" in 1643 for the description of internal organs reversed position in a patient. The first operation of coronary artery bypass grafting in a patient with dextrocardia was performed and described by Irvin RG., *et al.* in 1980 [3,4].

Case Report

Patient A., aged 62, with the diagnosis of CAD, PICS (2011) was admitted to the department of CAD surgical treatment and

minimally invasive coronary surgery of A.N. Bakulev NMRC CVS. The patient underwent PCI with RCA stenting in 2011 and 2017. Angina pectoris 3 FC. CD 2 A degree. CHF NYHA 2 FC. Dextrocardia. Hypertension 2 stage, 3 risk of cardiovascular complications. Chronic tubulo-interstitial nephritis. Hypertensive nephrosclerosis (GFR 41 ml/min/1.73 м2); CKD 3 B st. hyperuricemia. Non-acute chronic smoker bronchitis. Chronic erosive gastritis, remission.

According to the patient's history, he has been suffering for a long time from arterial hypertension with max 160/100 mm Hg, adapted to 120/80 mm Hg. The patient was diagnosed with dextrocardia (situs inversus) during army conscription commission. CAD manifested in 2008 by angina pectoris presentation. He was followed as an outpatient at his place of residence. The patient received all the time cardiotropic and antihypertensive therapy. He had left ventricle (LV) posterior wall acute myocardial infarction (AMI) in 2011, on this account coronary angiography (CA) was performed and followed with percutaneous intervention (PI) alongside with right coronary artery (RCA) stenting achieving satisfactory angiographic and clinical effect. The patient felt quite well later on, he regularly took prescribed drugs. In April 2017, he experienced angina again, underwent on this account second coronary angiography with single-step PI with RCA stenting. He was then followed as an outpatient at his place of residence. The patient started feeling worse in March 2021, angina symptoms progressed gradually with decreased exercise tolerance. He was admitted to the hospital at his place of residence in November 2021, where he underwent coronary angiography. CA results showed RCA dominance. Left main coronary artery (LCA) had no changes. Anterior interventricular artery (AIVA) had 85-90% stenosis in the middle third. Diagonal artery (DA) had up to 90% stenosis in the proximal third. Circumflex branch (CB) was occluded in the middle third, the distal third was filled through collaterals. Obtuse marginal branch (OMB) had 75-80% ostium stenosis, no changes further on. RCA was occluded in the middle third, the distal third was filled through collaterals. Posterior interventricular branch from right coronary artery (PIVB RCA) and posterolateral branch from right coronary artery (PLB RCA) were filled through collaterals. SYNTAX Score II was 13,6 (Figure 1-6).

Data obtained during patient's examination at the department: ECG: sinus rhythm, normal, with heart rate 61 bpm. No cicatricial or ischemic lesions were observed.

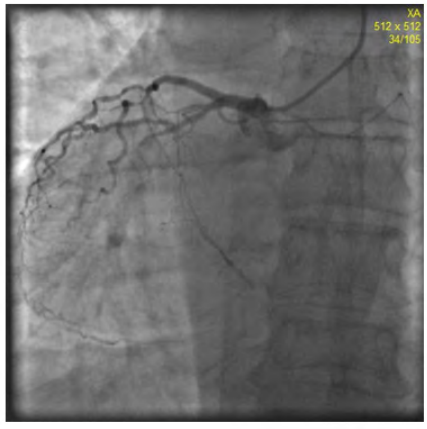

Figure 1: LCA.

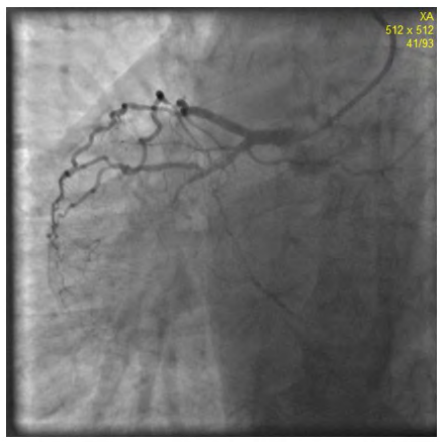

Figure 2: LCA 2.

Figure 3: LCA 3.

Figure 4: LCA 4.

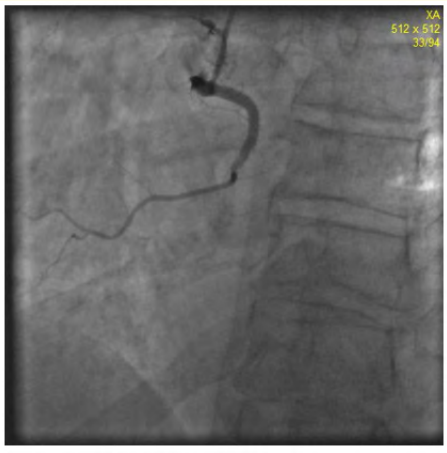

Figure 5: RCA.

Figure 6: RCA 2.

LV hypertrophy. Echocardiography: heart is located in the right part of thoracic cavity with mirror position of cavities and main vessels. Ascending aorta 37 mm, left atrium (LA) apically 46 x 57 mm, right ventricle (RV) 3,4 cm, end systolic volume (ESV) 41.0 ml, end diastolic volume (EDV) 94.0 ml; stroke volume (SV) 53.0 ml; LV ejection fraction (EF) 56.4%, thickness of the interventricular septum (TIVS) 13 mm, LV posterior wall thickness (LVPWT) 12 mm, right atrium (RA) 40 mm. Signs of initial pulmonary hypertension were observed - systolic pressure in the pulmonary artery 35 mm Hg. Mitral (MV) and tricuspid valve (TV) insufficiency of 1,5 degree.

Multislice computed tomography (MSCT) data: Heart: left-formed, right heart, enlarged moderately. The heart structures are mirrored. The pericardium with clear, even contours, the fluid in its cavity is not defined. Aortic valve: tricuspid, leaflets thickened. Coronary arteries: originate mirror-like, walls with combined atherosclerotic plaques (ASP) with calcium inclusions. The stent is viewed in RCA middle third. Vena cava are located on the left, go into RA mirror-like: SVC 21x23 mm, IVC - 25x28 mm. Atria: RA 50x55mm, LA up to 41x53x68 mm. Pulmonary veins are drained into left atrium. On the right PV end by a common trunk into the LA, diameter 25 mm, length 26 mm. Ventricles: RV - cavity dimensions during diastole (anterior-posterior dimension at the middle) up to 45 mm; LV - cavity dimensions during diastole at the middle 61 mm. LVOT up to 27 mm, interventricular septum thickness during diastole at the middle (at average diastole) up to 10 mm. VSD is not detected.

Aorta originates mirror-like from LV: AAo at the level of FR 20x26 mm, at the level of Valsalva sinuses - 38 mm, at the level of sinotubular junction - 30 mm, at the level of PT bifurcation 36 mm. Ao arch is right, at the cross-section 29 mm. Descending aorta is located to the right of the spine, at the level of PA trunk - 26 mm, at the level of LA - 25 mm, at the level of diaphragm 24,5 mm. BCA: originate mirror-like from Ao arch, BCT and RCCA originate with common trunk with the diameter of 16 mm, right subclavian artery - 12 mm. Pulmonary artery originates from RV, located mirror-like on the right: PT is not dilatated, diameter 25 mm, RPA - 23 mm, LPA - 23 mm (Figure 7,8). According to the examination level: situs inversus of abdominal organs (liver, spleen, stomach).

Figure 7: MSCT.

Figure 8: MSCT.

Chest X-ray: Heart dextraposition

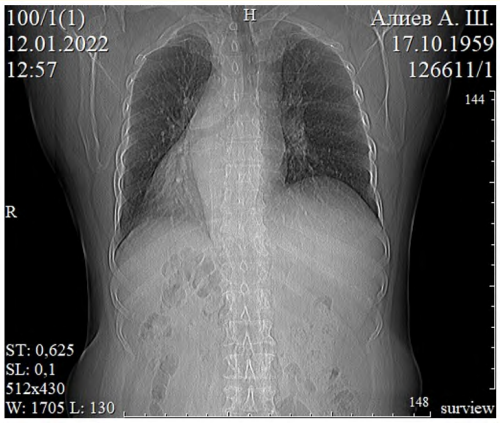

Figure 9: Chest X-ray.

Direct myocardial revascularization was performed 17 January 2022. It included mammary coronary bypass by anterior interventricular artery, coronary artery bypass grafting by autogenous grafts of diagonal artery and obtuse marginal branch, coronary artery sequential bypass grafting by autogenous graft of acute marginal branch and right coronary artery under parallel perfusion and normothermia. Total cardiopulmonary bypass (CPB) time was 168 min (Figure 10).

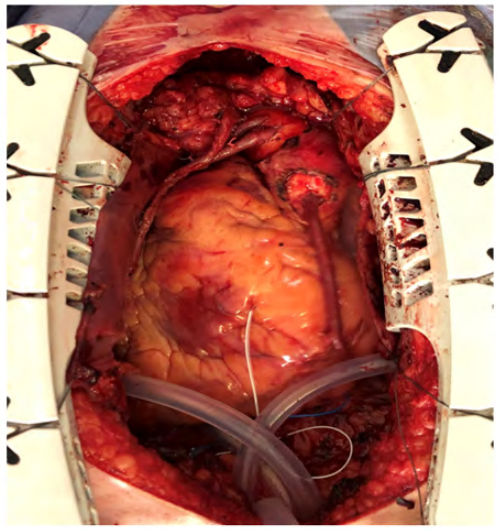

Figure 10: The final stage of the operation.

Bypass angiography was performed intraoperatively: bypasses are patent all through, anastomoses are secure (Figure 11-14).

Figure 11: RITA - AIVB.

Figure 12: CABG OMB.

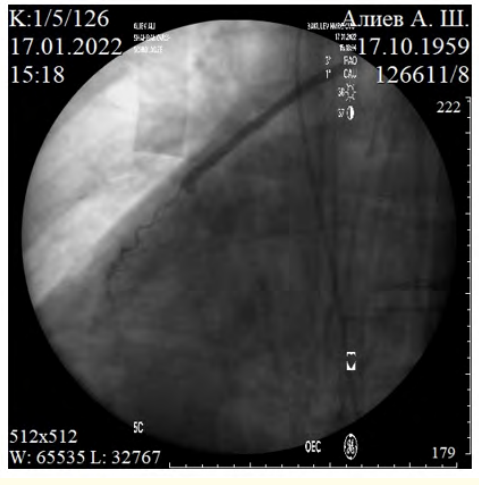

Figure 13: CABG DB.

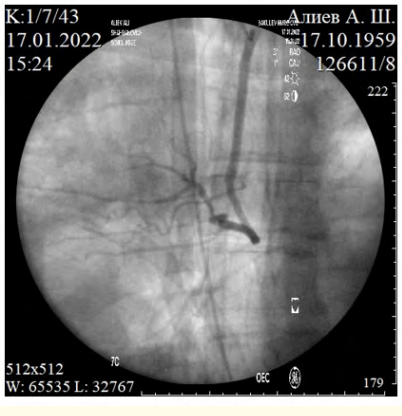

Figure 14: CABG PLA RCA and AMB.

The patient was transferred to intensive care unit. Mechanical ventilation lasted for 7.5 hours. The patient was transferred to the department the next day. Early postoperative period was fine, no complications were observed. Haemodynamics was stable. Postoperative clinical data as well as instrumental and laboratory tests results related to the severity of operation which the patient underwent. EchoCG data demonstrated no negative changes: LA - 40 mm; ESV 39.0 ml EDV 92.0 ml SV 53.0 ml EF 57.6%. Heart cavities were not dilatated. LV myocardium contractility was satisfactory. There were no signs of free fluid in pericardium and pleural cavities. Chest X-ray showed that the lungs were expanded. There was no free fluid in pleural cavities. Heart sizes were stable. The patient was discharged on the 10th day after the operation in a good condition.

Discussion

Dextrocardia is an anomaly associated with gene defect; HAND, ZIC3Shh, ACVR2, Pitxz gene mutations which develop because of disorders of embryogenesis processes at the early stages of intrauterine development. Situs viscerum inversus totalis means total or partial mirror-like position of organs in chest and abdomen. Dextrocardia is one of transposition variants which was first described by an Italian surgeon and anatomist Marco Aurelio Severino in 1643 [5-8].

There are 4 types of transposition:

- Situs inversus cordis — simple dextrocardia, only heart is located atypically in the right of the thorax;
- Situs viscerum inversus partialis — partial inversion, heart and some organs of gastrointestinal tract and respiratory system are mirrored;
- Situs viscerum inversus totalis — all gastrointestinal tract and respiratory system organs are mirrored;
- Situs ambiguous (heterotaxy) — a rare variant with ambiguous location, liver is usually in the middle of the body, spleen is absent.

The patient in our case had 3 type (situs viscerum inversus totalis). During intraoperative exploration left-formed, right heart was found. Taking into account heart transposition, right internal thoracic artery (RITA) was used as a conduit for anterior interventricular artery (AIVA), autogenous anastomoses were used as well.

There is a limited number of reports about myocardium revascularization in such patients in the literature [9].

Dextrocardia is a rare congenital malformation of the cardiovascular system, in which the heart is located in the chest on the right, in a manner similar to the normal location. Separate descriptions of surgical treatment of ischemic heart disease in patients with SIT are given in the relevant literature. Most surgeons in these operations stand on the left side of the patient [3]. Saad RA at all conducted an analysis of the available literature. The question addressed was which side of the operating table you should stand on when carrying out surgical revascularization on a patient with dextrocardia. Altogether 40 papers were found using the reported search, of which 19 represented the best evidence to answer the clinical question. The authors, journal, date and country of publication, patient group studied, study type, relevant outcomes, and results of these papers are tabulated. The side on which the operating surgeon stood was mentioned in 20 out of the 24 cases. Surgery was carried out from the conventional right side of the patient in 5 cases, while in 10 cases, it was carried out from the left side. The surgeon needed to switch sides to facilitate surgery in three cases. In addition, the right internal mammary artery (RIMA) was anastomosed to the left anterior descending artery (LAD) in 16 cases. Of these, surgery was carried out from the left side in 11 cases. The left internal mammary artery (LIMA) to LAD anastomosis was carried out in two cases, one of which was a free LIMA graft. In six cases, only vein grafts were used. Fourteen cases were carried out using cardiopulmonary bypass while 10 cases were carried out as off-pump cases with one conversion. The majority of patients were operated on from the left of the table. More cases were performed with the RIMA as the conduit of choice to the LAD [10].

This operation can be performed either on off-pump or on-pump. Murtuza B., *et al*. We analyzed 32 cases of patients with SIT who underwent CABG surgery in the period from 1981 to 2009. A total of 20 patients underwent CABG on-pump, 11 off-pump (one patient had a conversion, i.e. switching to a heart—lung machine during surgery) and in one case it was not reported. Large subcutaneous veins were used as conduits in 6 cases, arterial grafts in 7 cases, and both types of grafts were used in 19 cases. The exposure of surgical targets for CABG in dextrocardia patients may be difficult with a conventional operative arrangement. Most reported operations were performed while the surgeon stood on the left side of the operating table. This position facilitates grafting to vessels in both coronary artery distributions and particularly to the circumflex region, which is exposed by retracting the heart toward the surgeon on the left side of the table. The LAD anastomosis can probably be performed with similar ease from either side of the table. Because the right ventricle (RV) is on the left and anterior to the LV, exposure of the right coronary branches is perhaps easier from the left side. If the surgeon stands on the left side, the position of the stabilizer/retractor device for OPCAB grafting may also need to be modified [8,11-14].

The anatomy of this condition requires few adjustments during the conduct of surgery. In dextrocardia, the surgeon will operate from the left side of the patient in most centers. However, surgery can also be performed by the surgeon in the conventional position [15].

Despite the fact that most surgeons prefer to stand to the left of the patient when applying an anastomosis in patients with dextrocardia due to the position of the heart. However, there are reports showing that surgeons operate in a normal position. The presence of a bilateral superior vena cava should be checked before vein catheterization. Ravikumar MS and all showed - RIMA is usually harvested instead of the left internal mammary artery (LIMA) in these cases. In our case, the only time the surgeon positioned himself to the left of the patient was while harvesting RIMA. LIMA is usually avoided since the length of the graft will not be adequate to anastomose to the LAD resulting in the stretch of the graft and inadequate flow. The LIMA will have to cross the midline exposing it to high chances of injury during the re-do surgery. The LIMA flow may get compromised due to marked angulation at its origin leading to a dismal result of surgery. These limitations are avoided by using RIMA. Other considerations before incision include placement of electrocardiogram electrodes over the right hemithorax and placement of a central venous catheter in the left-sided internal jugular vein which will be monitored by the cardiac anesthetist [15].

The majority of surgeons when operating a patient with dextrocardia prefer to stand next to the left side of the patient due to the heart position. In our case, the surgical team stood contralaterally as well. According to the majority of authors [3,8], surgeon's position next to the left side of the patient during the intervention is the best in such patients.

Conclusion

The described clinical case is interesting because of a rare congenital anomaly. The surgical team must have certain skills to conduct coronary artery bypass grafting in a patient with dextrocardia and SIT. Surgeons face some problems and questions: patient's preoperative preparation, surgeon's position at the left or at the right during the operation, conduit choice for anastomosis. This particular clinical case presents research interest because dextrocardia combined with situs viscerum inversus totalis are rarely met. In our opinion, surgeon's position next to the left side of the patient during the operation is the best in such patients. Besides, it is more preferable to use right internal thoracic artery for creating anastomosis with anterior descending artery. Thorough patient's examination, taking into account different types of dextrocardia, is necessary. Coronary artery bypass grafting can be successfully performed in patients with dextrocardia.

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