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oronary Artery Bypass Grafting in a Patient with Congenital Chest Abnormality with Poland Syndrome

Michael Mikhailovich Alshibaya [1], Maxim Leonidovich Mamalyga [1,*], Dzhanybek Yerikovich Musin [1], Alexey Vladimirovich Dorofeev [2], Gela Guramovich Getsadze [2], Konstantin Vladislavovich Krymov [1] and Zurab Merabovich Cheishvili [1]

[1]Department of Surgical Treatment of Ischemic Heart Disease, A.N. Bakulev National Medical Research Center for Cardiovascular Surgery, Moscow, Russian Federation
[2]Department of Radiation Diagnostics, A.N. Bakulev National Medical Research Center for Cardiovascular Surgery, Moscow, Russian Federation

[*]*Corresponding author*: Department of Surgical Treatment of Ischemic Heart Disease, A.N. Bakulev National Medical Research Center for Cardiovascular Surgery, Moscow, Russian Federation. Email: mamalyga83@mail.ru

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Abstract

Introduction: A rare clinical case of coronary artery bypass grafting in congenital chest abnormality with Poland syndrome is described. This syndrome is a congenital anomaly, which is characterized by a combination of unilateral aplasia or hypoplasia of the sternocostal head of the pectoralis major muscle with ipsilateral brachysyndactyly in the classic version. The cause of Poland syndrome is unknown, but it is believed that it can form with a short-term decrease in blood flow in the subclavian and vertebral arteries.

Case Presentation: The presented case shows the possibility of successful completion of coronary artery bypass grafting in Poland syndrome. Revascularization by transluminal balloon angioplasty with stenting reduces the risk of postoperative complications. However, given the degree of damage to the coronary bed and myocardium, in some cases, the patient needs to undergo only coronary artery bypass grafting. The presented case shows the possibility of successful completion of coronary artery bypass grafting in Poland syndrome.

Conclusions: We believe that, despite the high risks of postoperative complications, patients with Poland syndrome can be recommended to perform a planned operation of coronary artery bypass grafting, including access through a median sternotomy.

Keywords: Coronary Artery Bypass Grafting, Poland Syndrome, Thoracotomy

1. Introduction

Poland syndrome was first described in 1840 by medical student Alfred Poland. This syndrome is a rare congenital anomaly, which is characterized by a combination of unilateral aplasia or hypoplasia of the sternocostal head of the pectoralis major muscle with ipsilateral brachysyndactyly coronary bed in the classic version. In Poland syndrome, there are malformations of the anterior chest wall and breasts. In rare cases, dextrocardia, malformations of the kidneys, vertebrae and lower extremities are described (1-3).

The incidence rate is 1: 30,000 and is more commonly reported in men (1). In 75% of cases, unilateral disorders are observed in the right half of the chest (4). Bilateral agenesis of the pectoral muscle is very rare (1, 5). Most cases of Poland syndrome are reported sporadically. The mechanism of paradominant inheritance is known, in which mutations can be transmitted through many generations in the absence of an explicit phenotype (5).

The cause of Poland syndrome is unknown, but it is believed that it can form with a short-term decrease in blood flow in the subclavian and vertebral arteries (6, 7). As a rule, this diagnosis can be made only after the birth of a child (8). As a result of funnel-shaped chest deformity, patients with Poland syndrome often have pronounced changes in pulmonary function and cardiac disorders, which are caused by displacement and rotation of the heart around the longitudinal axis, overload of the right heart and expansion of the aorta (9).

Patients with significant deformities of the chest wall may require surgical reconstruction, which is performed after the completion of growth (1). During reconstructive procedures, we use different variants of costal autotransplantation in the first stage and the transposition of the latissimus dorsi into the position of the large pectoral muscle in the second stage to complete the bone defect.

The objective of this report is to present a rare clinical case of successful CABG surgery in a patient with Poland syndrome.

2. Case Presentation

A 56-year-old gentleman was admitted to the A.N. Bakulev National Medical Research Center for Cardiovascular Surgery with the diagnosis: (1) atherosclerosis; (2) CHD; (3) effort angina 3 FC; (4) chronic heart failure NYHA FC II; (5) condition after PCI - stenting of the right coronary artery (06.05.2018); (6) hypertension of the grade III; (7) chronic mixed gastritis; (8) congenital deformity of the chest.

According to the coronary history, angina attacks in the patient first occurred in 2016. In May 2018, the patient suffered a lower-localization myocardial infarction, followed by right coronary artery stenting. Due to the underlying ailment, the patient's general condition was moderately severe at the time of admission. Objective examination determines funnel-shaped deformity of the chest; right-sided deviation of the sternum; scoliosis; right-sided aplasia of the smaller and greater pectoral muscles; absence of costal cartilage of the II, III ribs, as well as shortening of the V, VI ribs.

According to the data of coronary angiography, the patient revealed stenosis of the anterior interventricular branch in the proximal part up to 70%; the circumflex artery (CX) in the proximal third was occluded; the right coronary artery (RCA) was diffusely altered throughout, stenosed in the proximal third up to 60%, in the middle third there was a stent with signs of restenosis up to 90%, and in the distal third there was stenosis up to 75 % (Figure 1).

Echocardiographic examination: aortic root diameter 35 mm, ascending part of the aorta 34 mm, left heart enlarged, left atrium 42 × 50 mm; marginal aortic valve fibrosis, fibrous ring 23 mm, peak gradient 9 mm Hg; left ventricle - end systolic volume 114 mL, end diastolic volume 187 mL, stroke volume 73 ml, EF 39 %, akinesia of the basal, middle segments of the posterior and posterolateral walls, hypokinesia of the basal, middle segments of the posterior interventricular septum and anterolateral wall; mitral valve - cusps fibrosis, mitral-papillary dysfunction, fibrous ring 37 mm, peak pressure gradient 3 mm Hg, regurgitation 2.5 degrees; fibrous ring of the tricuspid valve 30 mm, grade 1 regurgitation, systolic pressure in the pulmonary artery 27 mm Hg.

The electrocardiographic study recorded sinus rhythm with a frequency of 70 per minute, Q-waves in leads III, aVF, ST segment was isoelectric.

According to the X-ray examination of the chest organs, the lungs are straightened, focal-infiltrative changes are not detected, paresis of the right dome of the diaphragm, thickened costal pleura on the right, paracostal fluid is determined in the right pleural cavity, dimensions 1 × 3.5 cm, the heart is shifted to the left, moderately enlarged, deformity of the chest on the right at the level of the angle of the scapula is apparent.

2.1. Computed Tomography

the chest is funnel-shaped deformed, an anomaly of the ribs on the right is visualized, the absence of costal cartilage of the II, III ribs, shortening of the V, VI ribs (hypoplasia, torn thinned anterior segments with curvature); the second rib has an abnormal attachment to the sternal handle; right-sided deviation of the sternum; curvature of the axis of the thoracic spine, the angle of inclination to the right, the vertebral bodies are rotated (Figure 2) aplasia of the smaller pectoral muscles and greater pectoral muscles, reduction of the thickness of subcutaneous fat on the right. The heart is located typically and its size is moderately increased. The roots of the lungs are not expanded. The lung pattern is not changed. No foci of consolidation were identified. No fluid is detected in the pleural cavity.

Based on the data of an objective examination, X-ray and computed tomography of the chest organs in the A.N. Bakulev National Medical Research Center for Cardiovascular Surgery, the patient was given a rare diagnosis of Poland syndrome.

According to external respiration data, the patient has moderate restrictive disorders. Values of bronchial patency are not altered.

Taking into account the clinical picture of angina pectoris, multivessel damage of the coronary bed (proximal prolonged stenosis of the anterior interventricular branch up to 70%, occlusion of the Cx, stenosis of the right coronary artery up to 90% distal to the previously implanted stent), the patient is shown to perform coronary bypass surgery.

The patient underwent the operation of aortocoronary bypass surgery of the anterior interventricular branch and posterior interventricular branch of right coronary artery with autoveins. Operation conditions: median sternotomy, parallel artificial blood circulation. Artificial blood circulation time was 60 min. The choice of surgical approach was determined by the need to perform revascularization of the anterior interventricular branch and the critically affected right coronary artery. Such revascularization is technically difficult to perform with a left-sided lateral thoracotomy. Due to the high risk of damage to the hypoplasticized sternum and potential instability in the postoperative period, the isolation of the left in-

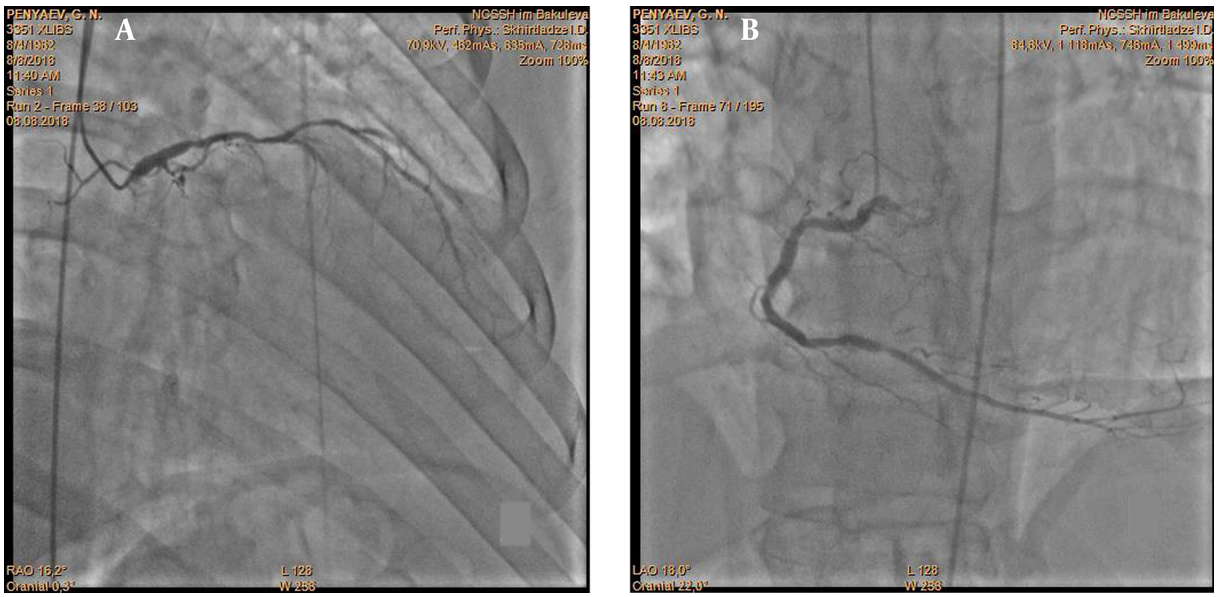

Figure 1. Coronary angiography of the A, left; and B, right coronary arteries.

ternal thoracic artery was not performed. Great saphenous vein is isolated on the right lower limb. During the operation, an increase in the size of the heart and extensive scarring of the posterolateral wall of the LV were revealed. Circumflex branch and obtuse marginal branch are diffusely atherosclerotic throughout. Autovenous bypass grafting was performed for the posterior interventricular branch of right coronary artery distal to the stent and the anterior interventricular branch in the middle third. We had a standard end of artificial blood circulation. The sternum was sutured with four wire stitches (Figure 2).

The early postoperative period was smooth. The patient was extubated on day 1 after the surgical intervention. In postoperative computed tomography, the right-sided deviation of the sternum is visualized, which is fixed by four wire sutures, the integrity of the sutures is not violated; the mediastinum is not displaced nor expanded. There are no focal changes in the lungs. Fluid in the pleural cavities is not detected. According to the ECHO CG, which was performed on the 6th day after the operation, LV end systolic volume - 105 mL, LV end diastolic volume - 181 mL, LV stroke volume - 76 mL, LV EF - 42 %, up to grade 2 mitral valve regurgitation and there was no pericardial effusion. The patient was discharged from the hospital on day 11 in a satisfactory condition.

3. Discussion

Thoracic interventions in patients with congenital funnel-shaped chest abnormality in Poland syndrome are associated with a high risk of developing respiratory failure in the postoperative period and potential wound complications. According to the literature, there are many sources describing open cardiac surgery for this pathology in children with congenital heart defects (both radical and palliative), accesses and median sternotomy and lateral thoracotomy are used. Revascularization by transluminal balloon angioplasty with stenting could significantly reduce the risks of postoperative complications, but given the degree of damage to the coronary bed and myocardium, in this case, the patient needed to undergo only coronary artery bypass grafting. It should be noted that the choice of surgical access and the provision of the conditions of the surgical aid, in our opinion, is exclusively the prerogative of the specific clinic and the surgeon performing coronary bypass surgery. The anatomical polymorphism of Poland's syndrome probably leaves the possibility of all options for surgical treatment of CHD.

3.1. Conclusions

The presented case shows the possibility of successful completion of coronary artery bypass grafting in Poland syndrome. We believe that, despite the high risks of postoperative complications, patients with Poland syndrome

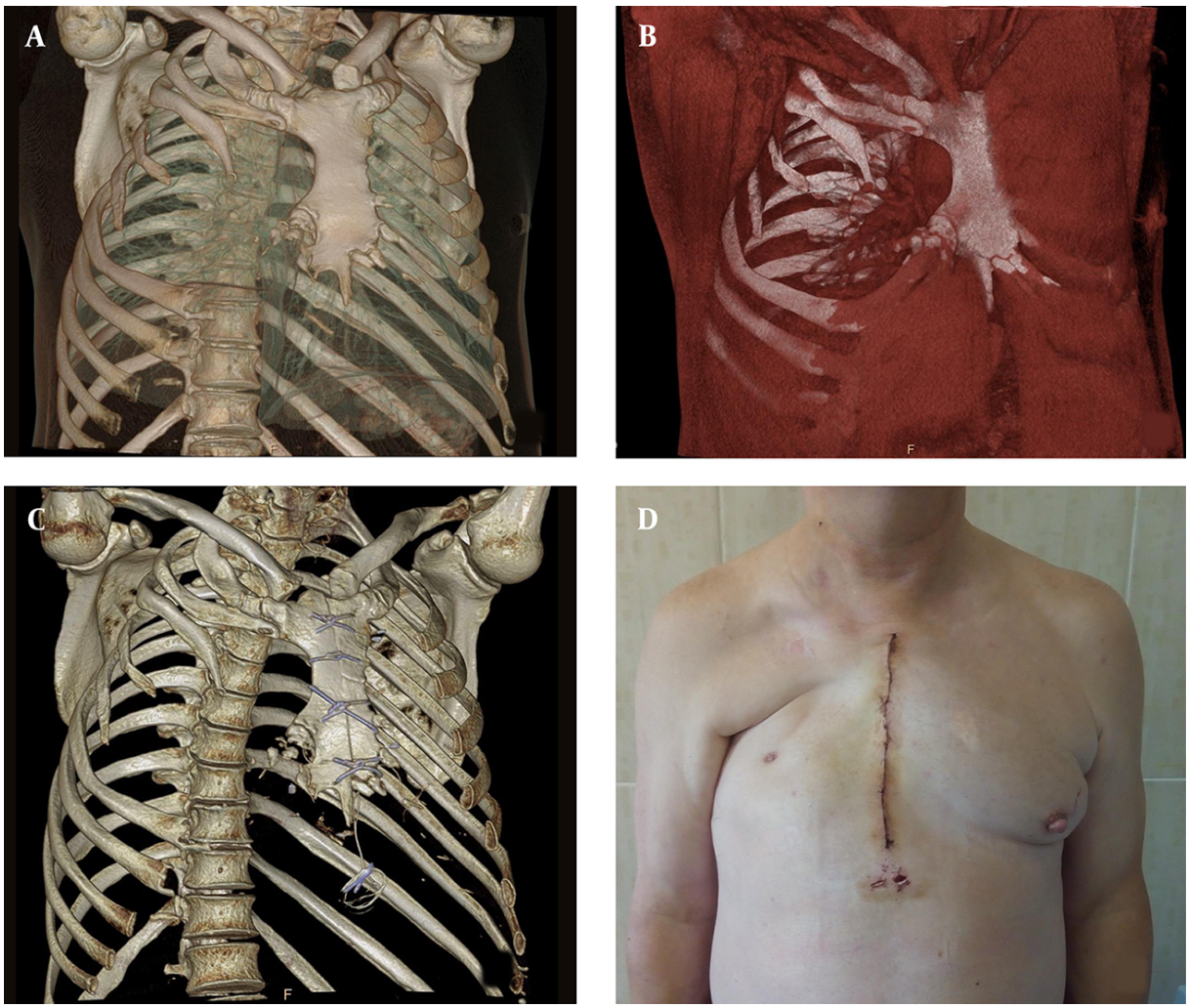

Figure 2. Computed tomography of the chest A, B, before; and C, after surgery; D, the appearance of the chest after surgery.

can be candidates to perform a planned operation of coronary artery bypass grafting, including access through a median sternotomy.

Footnotes

Authors' Contribution: Writing the paper, Alshibaya Mikhail Mikhailovich, Mamalyga Maxim Leonidovich; CT examinations and editing the manuscript, Dorofeev Alexey Vladimirovich, Getsadze Gela Guramovich; Performing surgery and editing the manuscript, Musin Dzhanybek Yerikovich, Krymov Konstantin Vladislavovich; Data gathering and helping with the writing of this article, Cheishvili Zurab Merabovich.

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