Case Report

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Neoaortic Bicuspid Valve Replacement in Patient After Arterial Switch Operation to Correct Dextro-Transposition of the Great Arteries

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INTRODUCTION Transposition of the great arteries is the second most common cyanotic congenital heart defect after tetralogy of Fallot. The arterial switch procedure (A. Jatene, 1975) is the surgical treatment of choice. Neoaortic root dilatation and valve regurgitation are quite common among the patients who underwent surgery for transposition of the great arteries. However, there are a lot of conflicting data about their direct connection.

CLINICAL CASE This article describes surgical repair of neoaortic bicuspid valve regurgitation, by it successful implantation, in an 18-year-old patient after arterial switch operation for transposition of the great arteries in the neonatal period.

CONCLUSION Neoaortic valve insufficiency can develop primarily as well as secondary to neoaortic root dilatation, however, the questions about valve repair or aortic root replacement with or without neoaortic valve implantation remains debatable. At this stage, decision making is based only on unsystematic clinical experience, surgeon's intuition, the basics of anatomy and pathophysiology, as well as close interaction of "pediatric" and "adult" cardiac surgeons.

Keywords: aortic valve, transposition of the great arteries, arterial switch operation, neoaortic valve, aortic regurgitation, heart valve replacement

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AV - aortic valve
HLM - heart-lung machine
CHD - congenital heart disease
BAV - bicuspid aortic valve
MV support - mechanical ventilation
EDV - end diastolic volume
ESV - end systolic volume
PA - pulmonary artery
LA - left atrium
IVS - interventricular septum
MV - mitral valve
MSCT - multislice computed tomography
NAVI - neoaortic valve insufficiency
RV - right ventricle
RA - right atrium
BSA - body surface area
SPPA - systolic pressure in the pulmonary artery
TV - tricuspid valve
TGA - transposition of the great arteries
EF - ejection fraction
CHF - chronic heart failure
HR - heart rate
EAH - electrical axis of the heart
EchoCG – echocardiography

Transposition of the great arteries (TGA) is the second most common cyanotic congenital heart disease (CHD) after tetralogy of Fallot. It amounts up to10% of all CHD, which corresponds to about 8 cases per 1000 live births [1]. The gold standard surgical treatment for TGA in newborns is arterial switch operation which was first performed on a patient with TGA and ventricular septal defect (VSD) by A. Jatene in 1975 [2]. Despite the seemingly anatomical repair of the defect, in which the risks of complications are supposed to be leveled, they still develop and more often depend on the variants of the anatomy of the main vessels and coronary arteries.

The most common complications of the late postoperative period are neopulmonary artery stenosis, neoaortic semilunar valve insufficiency, neoaortic root dilatation, and coronary heart disease [3]. It is also possible to develop sudden cardiac death, rhythm and conduction disorders, chronic heart failure (CHF) in earlier periods of life.

Neoaortic valve insufficiency develops in 7% of patients 10 years after arterial switch operation and in 35% of patients 5 years after arterial switch operation with elimination of left ventricular (LV) outflow tract obstruction [4, 5].

In case of bicuspid aortic valve (BAV), the spatial arrangement of the structures of the aortic root is disturbed. The leaflets of the aortic valve are formed as right and left or as anterior and posterior. These anatomical changes, due to uneven distribution of the pressure force on the aortic valve leaflets, lead to overloads of the previously mentioned structures and to the formation of aortic root dilatation and aortic insufficiency [6].

Neoaortic root dilatation is quite common among patients surgically treated for TGA, but, according to the literature, there are conflicting data on its direct relationship with the development of neoaortic valve insufficiency [7–10].

This report presents a clinical case of surgical treatment for neoaortic bicuspid valve insufficiency by its successful prosthesis in an 18-year-old patient after anatomical correction of TGA by arterial switch operation performed in the neonatal period.

Clinical observation

Patient E., 18 years old, was hospitalized at the Department of Emergency Cardiac Surgery, Auxiliary Circulation and Heart Transplantation on April 15, 2019 with complaints of shortness of breath on minimal exertion, inability to take a deep breath, reduced exercise tolerance.

It is known from the patient's medical history that immediately after birth, he was diagnosed with TGA in combination with bicuspid pulmonary valve, patent foramen ovale and patent ductus arteriosus. On October 20, 2000 at the A. N. Bakulev National Medical Research Center for Cardiovascular Surgery an operation was performed: anatomical correction of the transposition of the great arteries (Fig. 1). The postoperative period was uneventful.

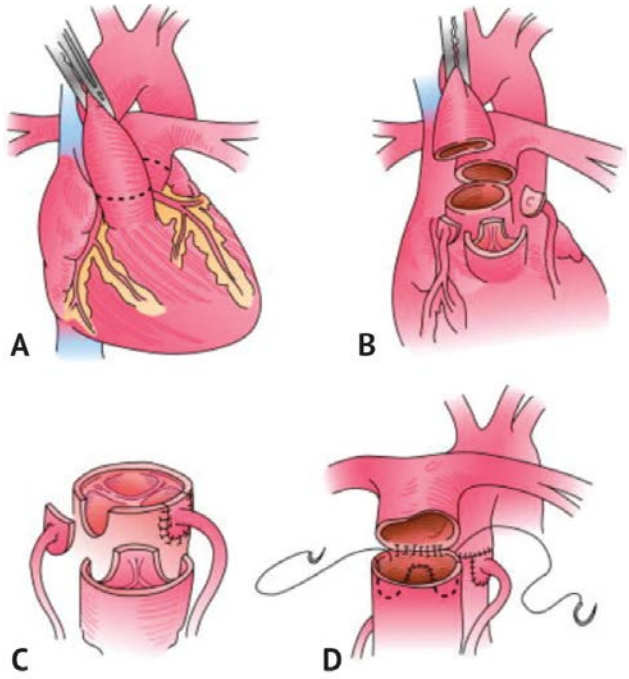

Fig. 1. Scheme of arterial switch operation: A — intersection of the great arteries; B — explantation of aortic sinuses' sections with the coronary arterial orifices; C — reimplantation of the coronary arteries into the neoaortic root; D — the Lecompte maneuver and restoration of the integrity of the ascending aorta and pulmonary trunk

On examination:

Electrocardiography: sinus rhythm with a heart rate of 52 per minute. Normal position of the electrical axis of the heart (EAH). No ischemic changes.

EchoCG: sinus part of the aorta 3.8 cm (Z-score 3.44), ascending aorta is not located, aortic arc 2.6 cm (Z-score 1.73). Left atrium (LA) 3.5 cm/50 ml (Z-score 1.93). Left ventricle (LV): end-diastolic volume/body surface area (EDV/BSA) 103 ml/m2, ejection fraction (EF) 52%, posterior wall thickness (PW) 1.0 cm, interventricular septum (IVS) 1.1 cm, local LV myocardial contractility is not impaired. Right atrium (RA) 40 ml. Right ventricle (RV) 2.5 cm. Aortic valve (AV): bicuspid, thickened leaflets with inclusions of calcium masses in the commissure area, no stenosis, grade 3 regurgitation into the LV cavity, AV fibrous ring 30 mm (Z-score 4 .54). Mitral valve (MV): leaflets are thin, no signs of stenosis, grade 1 regurgitation into the LA cavity, MV fibrous ring 32 mm (Z-score 0.59). Tricuspid valve (TC): leaflets are thin, no signs of stenosis, grade 1 regurgitation into the RA cavity. Systolic pressure in the pulmonary artery (SPPA) 21 mm Hg. (Fig. 2).

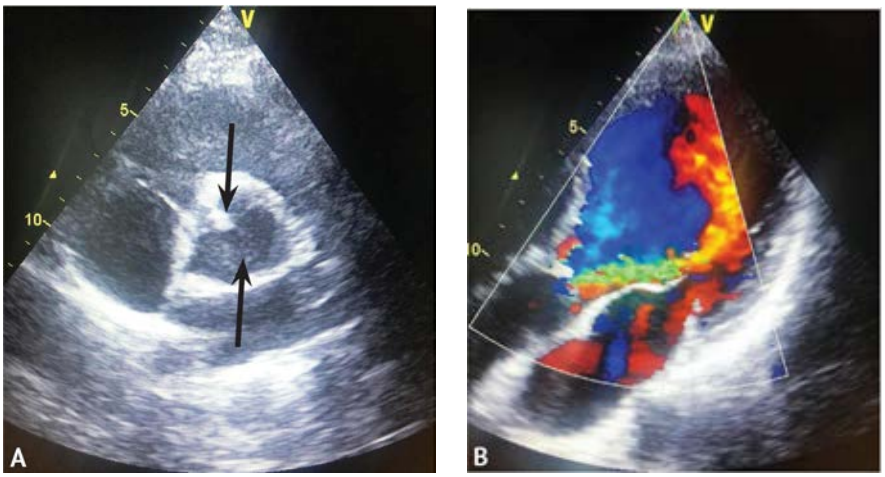

Fig. 2. Echocardiography before surgery: A — parasternal short-axis view. Two neoaortic valve cusps with calcium inclusions in the commissure region are visualized; B — apical 5-chamber view. Grade 3 aortic regurgitation

Chest X-ray image, frontal projection: the heart is expanded in diameter due to the left sections, the waist is underlined (Fig. 3).

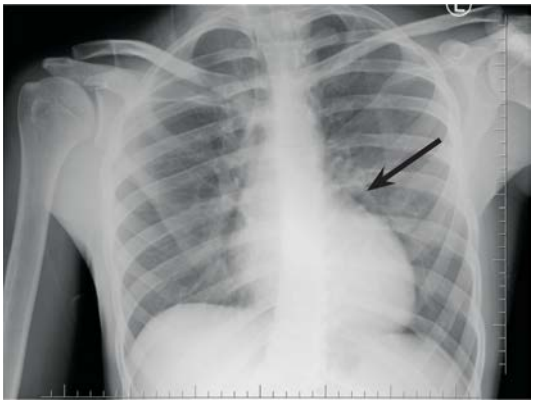
Fig. 3. Chest X-ray image, frontal projection. The radiograph has an appearance of an egg lying on its side, which is a sign of transposition of the great arteries and persists for life, despite the normalization of hemodynamics after arterial switch operation

Multislice computed tomography (MSCT) of the chest organs with contrast enhancement: thickening of the LV walls up to 12 mm is noted, the pulmonary artery (PA) trunk and its bifurcation are located anterior to the ascending aorta. In this case, the right branch of the PA is located between the ascending aorta and the superior vena cava. The diameter of the AV fibrous ring is 35 mm (Z-score 5.16). The diameter of the aortic root at the level of the sinuses of Valsalva is 43 mm (Z-score 3.9). The coronary arteries originate from the left and right facial sinuses of the aorta and have a typical epicardial course without signs of proximal stenosis. The ascending aorta 27 mm in diameter (Z-score 1.29), the aortic arch 22 mm (Z-score 0.69), the descending aorta 16 mm (Fig. 4).

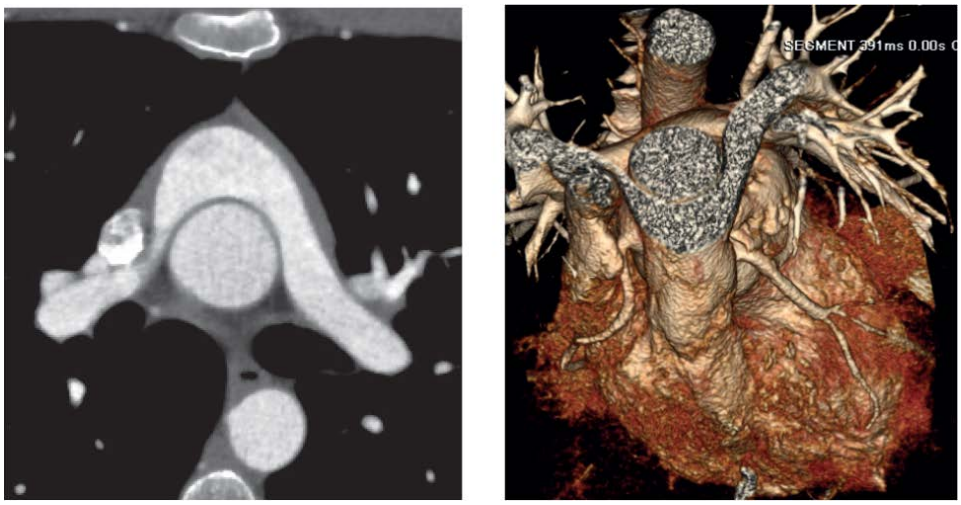

Fig. 4. Spiral CT with multiplanar and three-dimensional reconstructions. Division of the pulmonary trunk above the ascending aorta. The right and left pulmonary arteries go around the ascending aorta on both sides and occupy its entire anterior semicircle

Based on the results of the examination, it was concluded that the patient had LV volume overload due to severe AV insufficiency, a consequence of aortic root ectasia (Carpentier type I). Given the high Z-score only at the level of the aortic root (fibrous ring and sinus part, without the ascending aorta), a decision was made to minimize the volume of the operation.

On May 14, 2019, the patient underwent surgical treatment in the amount of On-X-23 mechanical valve implantation.

Features of the operation: Taking into account the anatomical location of the main vessels in order to minimize the risk of damage to the neopulmonary artery and the right heart, it was decided to perform resternotomy after connecting the heart-lung machine (HLM) with femoral artery-femoral vein cannulation. After partial cardiolysis, the superior vena cava was additionally cannulated to enhance venous return. Myocardial protection was carried out by retrograde injection of Custodiol solution into the coronary sinus. For clamping the aorta in a sharp way, the distal part of the ascending section was isolated directly above the bufurcation of the PA trunk. After aortic cross-clamping, access to the neoaortic valve was made by a longitudinal dissection of the anterior wall of the pulmonary trunk and then through longitudinal incision of the posterior wall of the pulmonary trunk and aorta soldered together. The aortic valve is bicuspid. After valve leaflet excision, a mechanical prosthesis was implanted according to the standard technique. The integrity of the aorta was restored along with the posterior wall of the pulmonary trunk. A twisted suture was placed on the anterior wall of the pulmonary trunk.

Cardiac activity at the end of the surgery recovered independently (sinus rhythm), hemodynamic stability was provided by norepinephrine infusion at a dose of 50 ng/kg/min. Cardiopulmonary bypass time was 133 minutes, aortic cross-clamping time was 80 minutes.

Hemodynamics in the postoperative period remained stable; by the end of the first day, cardiotonic support was completely abandoned.

The duration of mechanical ventilation (MV) support after surgery was 4 hours. The patient was transferred from the intensive care unit to a hospital department on the 2nd day.

On the 9th day after surgery, the patient was discharged from the hospital.

Control examinations were carried out immediately before the patient's discharge, as well as one year after surgical treatment. The dynamics of the results is presented in Table 1.

Table

The results of instrumental examinations of patient E

Methods and indicators of patient examination	Before surgery (baseline)	7 days after surgery	1 year after surgery
Echocardiography:			
Aortic root, cm	3.8	н/в	3,8
Left ventricular end diastolic volume, ml	180	140	118
Left ventricular end diastolic volume /body surface area, ml/m2	103	78	66
Left ventricular ejection fraction, %	52	50	61
Aortic valve regurgitation, grade	3	1	1
Peak prosthetic aortic valve gradient, mmHg	11	21	20
Mitral valve regurgitation, grade	1	1	0–1
Tricuspid valve regurgitation on the, grade	1	1	0–1
Pulmonary artery systolic pressure, mm Hg	21	18	18
NYHA functional class of chronic heart failure, class	III	II	0–I

When examining the patient before discharge, a decrease in LV volumetric characteristics (EDV: 140 ml, end systolic volume – ESV: 69 ml) with their continuing decrease in dynamics (after a year, EDV: 118 ml, ESV: 40 ml), and an increase in LV EF from 50 to 61% were noted.

Subjectively, the patient feels much better, does not describe a clinical picture of CHF, demonstrates good exercise tolerance (0 FC CHF according to NYHA). The patient is being monitored.

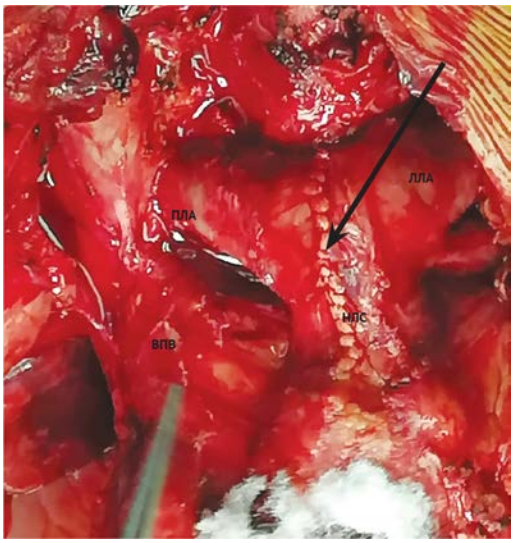

Fig. 5. Final view of the operation. The arrow indicates the site of access to the aortic valve (the suture of the anterior wall of the pulmonary trunk)

Notes: Ao — aorta; ВПВ — superior vena cava; ЛЛА — left pulmonary artery; НЛС — neopulmonary trunk; ПЛА — right pulmonary artery

DISCUSSION

Since the first successful TGA was performed by A. Jatene in 1975, survival rates have increased significantly due to the improvement of prenatal diagnosis, the possibility of using prostaglandins, performing surgical treatment in the first hours and days after birth, as well as the improvement of surgical techniques and capabilities of postoperative treatment [11].

One of the late complications after arterial switch operation is semilunar neoaortic valve insufficiency and neoaortic root dilatation.

According to some authors, the presence of an IVS defect, mild aortic regurgitation in the perioperative period, and a high ratio of the neoaortic root diameter to the ascending aorta are risk factors for NAVI development in the remote period after surgery [12].

The reason for the development of aortic valve insufficiency may be due to the fact that in its anatomical structure it is a pulmonary valve and is adapted for low pressure, which emphasizes the need for careful long-term monitoring and follow-up of the patient in order to identify risk factors in a timely manner and possibly avoid surgical interventions in some patients [8, 13].

The relevance of this problem is due to the lack of clinical recommendations, principles of evidence-based medicine and reliable data from clinical studies in the literature, which complicates decision making by the attending physician and the operating team.

It took the global community several decades of patient observation to gain experience before deciding on the need for reoperation for a patient with NAVI.

Although the overall rate of surgery for neoaortic dilatation and NAVI after TGA is still low (2.0–2.5%), some authors report that the development of regurgitation and dilatation is a time-dependent phenomenon requiring strict patient vigilance [9].

Despite the fact that a certain number of studies, statistical and clinical analyzes have already been carried out, data on reoperations are sporadic and further accumulation of clinical experience is needed.

According to the literature, NAVI of various degrees by the age of 20–23 occurs in 9.8–79.7% of cases [14]. In a study by van der Palen RLF et al., which included 345 patients who underwent anatomical correction of TGA in childhood, 47 patients developed the root diameter ⩾40 mm, and NAVI was present in 26% of them. A quarter of these patients have not even reached adulthood, and repeated surgical treatment was performed in 10 patients (2.9%) [10].

Ongoing neoaortic dilatation, progression of neoaortic valve regurgitation, and their relationship beyond childhood may predict an increase in future reoperations on the aortic root and valve [18]. However, by this age period, patients switch to treatment with “adult” cardiac surgeons, who rarely encounter pathology in newborns and childhood in their clinical practice.

The clinical picture in patients with BAV can vary from severe manifestations of valve dysfunction in infancy to their complete absence in the elderly [15]. Despite the variability of its clinical manifestations in an adult, the presence of this anatomical form of the valve structure is a significant risk factor for the development of aneurysm of the aortic root and ascending aorta.

In addition, the presence of BAV increases the risk of aortic dissection and rupture. Of greatest clinical significance is the type of BAV which occurs as fusion of one of the commissures (type 1 Sievers' valve classification), this is due to the low incidence of true BAV (type 0). The frequency of involvement in the pathological process of the aortic root or its tubular part differs depending on the anatomical orientation of the fused commissure, although the factors influencing this process remain not fully understood.

Surgical treatment of bicuspid aortic valve disease in adults in tactical terms does not cause any special discussions: unlike in children, the issue of tactics is solved much easier, since the vast majority of patients need aortic valve implantation and (or) aortic root with ascending aorta replacement [16].

In adult patients, the fact of isolated aortic valve repair for BAV does not lead to an increased risk of root or ascending aortic dilatation or aortic dissection in the long term. The use of valve sparing techniques for BAV in adults depends on the anatomy of the valve, the type of insufficiency, and the experience of the surgeon, and should always be considered in young patients [17].

CONCLUSION

Neoaortic valve insufficiency can develop primarily as well as secondary to neoaortic root dilatation, but the need for valve repair or aortic root replacement with or without neoaortic valve implantation remains debatable.

At this stage, clinical decision making is based only on non-systematized clinical experience, intuition of the attending physician and/or surgeon, knowledge of the basics of anatomy and pathophysiology, as well as close interaction of "pediatric" and "adult" cardiac surgeons.

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