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Historical and modern aspects of surgical treatment of Ebstein's anomaly

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ABSTRACT

Congenital heart defects (CHDs) are recognized as the most common type of congenital pathology. The frequency of CHDs reaches 2.4–14.2 % per 1000 newborns. Ebstein's anomaly is a rare and complex pathology that can be manifested clinically at any age. Drug treatment is ineffective in patients with this pathology. Preference in this case should be given to surgical treatment. Cardiac surgeons must know basic methods of correction of this pathology, their advantages, and disadvantages. The literature review shows the evolution of Ebstein's anomaly surgical correction techniques from the middle of the XX century to the present moment. The description of the main tricuspid valve repair techniques, which had an impact on the development of Ebstein's anomaly surgery, is given. The volume of flap tissue that can be separated from the wall of the right ventricle is the key to successful valve repair. A case of tricuspid valve repair and replacement is given. "Cone" reconstruction is the most promising modern technique. There is also a need in new techniques and modification of the existing ones. The works aimed at their improvement and elimination of imperfections are promising.

Key words: congenital heart defects, Ebstein's anomaly, tricuspid valve replacement, tricuspid valve repair.

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Исторические и современные аспекты хирургического лечения аномалии Эбштейна

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РЕЗЮМЕ

На сегодняшний день врожденные пороки сердца (ВПС) занимают одно из ведущих мест среди всей врожденной патологии. Частота ВПС достигает 2,4–14,2% на 1000 новорожденных. Аномалия Эбштейна – редкая и сложная патология, клиническая картина которой может проявляться в любом возрасте. Медикаментозное лечение пациентов с этой патологией малоэффективно. Предпочтение в данном случае следует отдать хирургическому лечению. Для кардиохирурга важно знать основные методы коррекции этой патологии, их преимущества и недостатки.

В литературном обзоре показана эволюция методов хирургических коррекции аномалии Эбштейна с середины XX в. по настоящий момент. Дано описание хирургической техники основных способов пластики трикуспидального клапана, оказавших влияние на развитие хирургии аномалии Эбштейна. Ключевым моментом для успешной пластики клапана становится объем ткани створки, который возможно отделить от стенки правого желудочка. Приведен опыт выполнения протезирования и пластики трехстворчатого клапана. Наиболее многообещающим методом на сегодняшний день является «конусная» реконструкция. Таким образом, существует необходимость поиска новых методов и модификации уже имеющихся. Работы, направленные на их усовершенствование и устранение недостатков, являются перспективными.

Ключевые слова: врожденные пороки сердца, аномалия Эбштейна, протезирование трикуспидального клапана, пластика трикуспидального клапана.

Конфликт интересов. Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

Источник финансирования. Авторы заявляют об отсутствии финансирования при проведении исследования.

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INTRODUCTION

Ebstein's anomaly (EA) is a rare congenital heart disease of the "blue" type, the incidence of which is 5.2 cases per 100,000 newborns, which is about 1% of all congenital heart defects [1-3]. Wilhelm Ebstein first described this pathology in 1866 during an autopsy of a nineteen-year-old patient, Josef Prescher, who died of chronic heart failure. Working as a physician's assistant and a prosecutor, he published his article describing this clinical case [4]. In our country, the first description of this pathology belongs to A.A. Elashevich (1925). The term "Ebstein's anomaly" as a designation of a nosological unit was first introduced into the literature in 1937 by Yater W. and Shapiro M., who described 16 clinical cases of this defect in their article [5]. According to various authors, the average life expectancy in the natural course of the disease is up to 50 years, with 80–87% of deaths occurring at the age of 30–40 years. In recent years, the interest of the surgical community specifically in plastic surgery in EA has increased, so the purpose of the literature review is to describe possible surgical approaches to correct this anomaly.

To date, there is no unequivocal opinion regarding the etiology of the disease. Most researchers are inclined to the multifactorial nature of this disease. Embryonic development of the tricuspid valve (TV) begins with the 5th week of intrauterine development. It has been established that precisely during this period disruption of the laying of the cardiac jelly between the ventricular myocardium and the endocardium occur, which leads to disruption of the delamination process, that is, embryonic separation of the tissue of the cusp from the tissue of the right ventricle (RV) [6].

EA is characterized by the following characteristics (Fig. 1) [7].

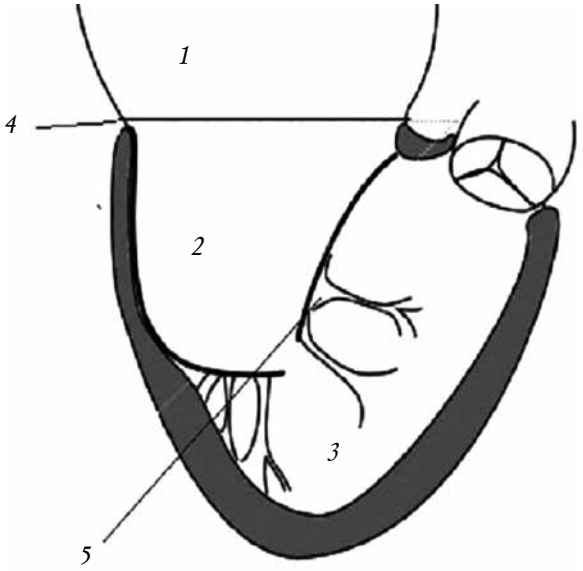

Fig. 1. Scheme of the structure of the right ventricle with Ebstein's anomaly: *1* – right atrium; *2* – atrialized part of the RV; *3* – the functional part of the RV; *4* – anatomical ring of the TV; *5* –functional ring of the TV

The first characteristic is the fusion of the cusps with the ventricular myocardium. As a rule, the anterior cusp is delaminated more from the ventricular wall, while the posterior and septal cusps are delaminated minimally. In the most severe cases, the septal cusp is a ridge of connective tissue. The second is the anterior-apical displacement of the functional ring of the TV (the place of transition of the atrialized part of the ventricle to the true right ventricle with normal delamination of the cusps) towards the outflow tract of the RV. The third characteristic is the redundancy of the tissue of the anterior cusp (sail-shaped cusp), its fenestration and limitation of mobility between the valve cusp and the RV due to chord connection. The fourth characteristic is the presence of a thinned, dilated and akinetic part of the RV (atrialized part), and the number of myocardial fibers in this area is much less than normal [8]. The fifth sign is the expansion of the true (anatomical) fibrous ring of the TV. As the severity spectrum increases, the fibrous transformation of the cusps from their muscle precursors increases during embryogenesis [9, 10]. In EA, the structure of the RV myocardium with a smaller number of myocyte fibers was altered, the nuclei of myocytes were displaced under the sarcolemma, and the Z-strips of sarcomeres were broken, so this anomaly can be considered a type of ventricular myopathy [11].

The most common concomitant defect is an atrial septal defect (42–60% of cases), while ventricular septal defect, transposition of the great vessels, and pulmonary stenosis are less common [10]. Due to the underdevelopment of the true fibrous ring, additional pathways function that are clinically manifested by Wolf – Parkinson – White syndrome in 7–30% of cases [12, 13].

In 1988, A. Carpentier [14, 15] (Fig. 2) proposed the following classification of Ebstein's anomaly.

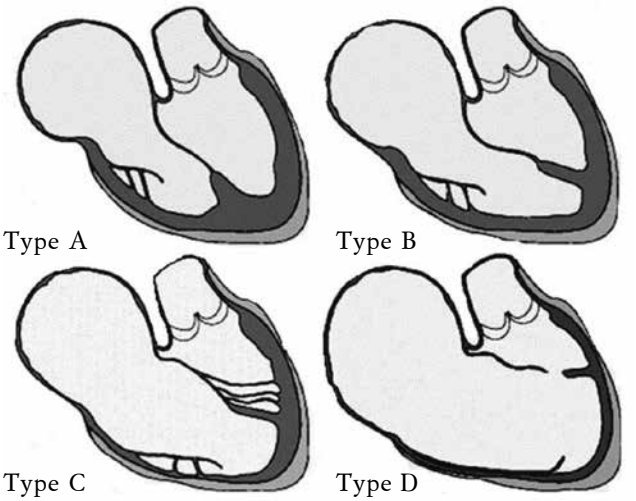

Fig. 2. Types of Ebstein's anomaly [14]: type A – true RV volume is satisfactory; type B – here is a large atrialized component of the RV, and the anterior cusp of the TV moves freely; type C – the anterior cusp of the TV is limited in its movement and can cause obstruction in the outflow tract of the RV; type D – almost complete atrialization of the ventricle with the exception of a small part of the outflow tract of the RV

In the Russian clinical guidelines, the extreme form of AE is additionally highlighted – type E: the fused anterior, posterior and septal valve cusps together with the inflow section of the RV form a "tricuspid sac". The wall of the inflow section of the RV is thinned. The communication between the "tricuspid sac" (atrium) and the infundibulum (ventricle) is often formed by a narrow hole in the region of the so-called anterior-septal commissure.

The natural clinical course of this pathology depends on the degree of TV dysplasia and function of the RV. Despite the extensive experience in surgical treatment, indications for surgery are not defined. To date, indications for surgical treatment are as follows: decreased exercise tolerance; cyanosis; progressive dilatation of the

right heart (cardiothoracic index over 60%); dysfunction of the right ventricle (ejection fraction below 30%, displacement of the interventricular septum towards the left ventricle); left ventricular dysfunction (decrease in ejection fraction below 50%, reduced end-diastolic volume); atrial arrhythmias.

Relative contraindications for correction according to J. Dearani et al. are [16]: age over 50; severe pulmonary hypertension; a significant decrease in left ventricular function (ejection fraction of less than 30%); a complete violation of the delamination of the septal and posterior cusps of the tricuspid valve, while the delamination of the anterior cusp is less than 50%.

But over the past few years, the approach to indications for surgical treatment has fundamentally changed. Since almost all patients require surgical intervention for this pathology sooner or later, the team of authors from the Mayo Clinic, led by J. Dearani, proposes to perform the operation as soon as the defect was diagnosed. This concept is based on the fact that with age, RV function deteriorates due to volume overload and the likelihood of right ventricular heart failure in the early postoperative period increases.

Ebstein's anomaly surgical correction options include:

1. Two-ventricular correction (valve prosthetics (biological or mechanical prosthesis), valve plastic).

2. One and a half ventricular correction (plastic or prosthetic valve with bi-directional cava pulmonary anastomosis).

3. One-ventricular correction (Starnes operation).

Until the middle of the last century, treatment of patients with EA was extremely conservative. The results of drug therapy were not significant; therefore, the main priority was for surgical care. Currently, all operations are performed on an open heart using a cardiopulmonary bypass and cardiac arrest.

Most babies in the newborn period do not require surgery. However, in extreme forms of the defect with severe clinical manifestations such as shortness of breath, cyanosis, and tachycardia and in the absence of the effect of prostaglandin E_1 therapy, surgical intervention is necessary. Right ventricular failure is also increased by the increased pulmonary vascular resistance, which persists in the first few weeks of life. To assess the survival of such patients in 1992, the GOSE scale (Great Ormond Street Echo) was developed [17]. The GOSE index is equal to the ratio of the sum of the sizes of the right atrium and the atrialized part to the sum of the left atrium, left ventricle and the functional part of the RV (the dimensions of the heart chambers are determined in the four-chamber position). Depending on the number of points, the risk of death in the natural course of the defect is determined.

Table 1

GOSE SCORE		
Parameter	SCORE, in points	Mortality, %
GOSE I	<0.5	0
GOSE II	0.5–0.99	10
GOSE III	1–1.49	44
GOSE IV	>1.5	100

If the anatomy of the cusps and ventricle is not suitable for performing two-ventricular correction, one-ventricular correction is performed (Fig. 3). This technique was first introduced by V.A. Starnes in 1991, 27 patients with the Starnes procedure in 1989–2015 were operated on at Children's Hospital Los Angeles.

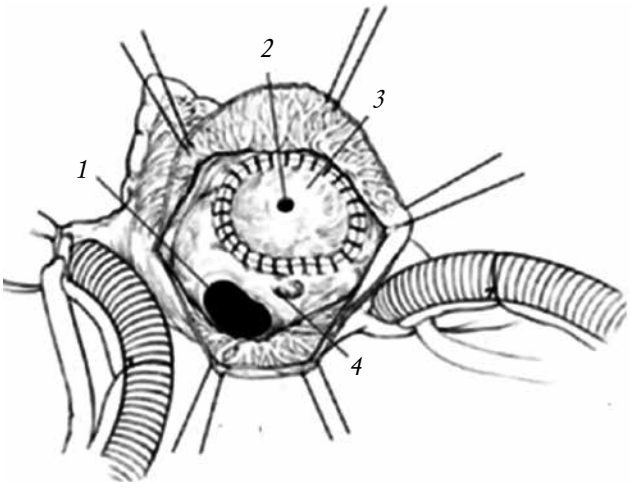

Fig. 3. Starnes operation. Image modified from [18]: *1* – dissected interatrial septum; *2* – fenestration PTFE patch 4 mm; *3* – PTFE patch; 4 – coronary sinus

An exception is made from the bloodstream of the RV by fixing a polytetrafluoroethylene patch with a fenestration of 4 mm to the true fibrous ring. In this case, the coronary sinus remained on the side of the right atrium. An atrial septum was additionally excised to form a single atrium. [18] Pulmonary blood flow was provid-

ed by a modified Blok – Taussig shunt. At the next stage of hemodynamic correction at the age of 3–6 months, a bi-directional cava-pulmonary anastomosis is performed; at 2–4 years old, total cava-pulmonary connection is performed. In newborns with an extreme form of Ebstein's anomally, the right ventricle is dilated and is not able to function adequately to maintain cardiac output; therefore, it is necessary to exclude the RV from the bloodstream [19, 20].

The surgeon is most interested in two-ventricular correction by means of plastic surgery or valve prosthetics. The first prosthetics of TV in EA was performed by C. Barnard in South Africa (Cape Town) in 1963. Then the procedure was modified by cardiac surgeons D. Ross and J. Somerville, who used an aortic homograft in 1970 to avoid the need for anticoagulants [21]. In 1988, N. Kumar and B. Dubey used pulmonary homograft for implantation in the tricuspid position. In the USSR, a successful valve replacement was performed by G.M. Soloviev in 1964 [22].

Valve prosthetics are possible using a biological or mechanical prosthesis [23]. A characteristic of prosthetics is that the valve is located above the true fibrous ring of the TV. The tissue of the cusps, causing obstruction of the outflow tract of the RV, must be excised, and the true fibrous ring is narrowed to the size of the prosthesis. The atrialized part of the RV is also reduced. On the posterolateral wall, the tissue is usually thinned, so the suture line should be closer to the atrium to avoid damage to the right coronary artery. To avoid damage to the atrioventricular node, the suture line is located above the coronary sinus, thus, the drainage of venous blood will be carried out in the RV.

Among the methods of surgical treatment, valve replacement for this pathology has significant disadvantages compared with reconstructive surgery. According to M. Brown et al., the long-term results of 378 TV prosthetics operations are: 6% mortality in the early postoperative period and 17% in the first ten years after. The frequency of prosthetics was 41% over the next 20 years [24].

In 2007, H. Bartlett et al. in their study, they described the results of TV prosthetics in 97 patients whose average age was (2.9 ± 1.7) years. 44 children received a mechanical prosthesis and 53 received a biological prosthesis. As a result, 26 (27%) patients died in the early postoperative period. Among the complications in the early postoperative period, complete atrioventricular blockade was observed in 13 patients, which required the implantation of a pacemaker. In the group with a mechanical prosthesis, the frequency of obstruction was higher than in the group with a biological prosthesis (23 versus 6%). Valve thrombosis in the early postoperative period was observed in five patients. All these patients received a mechanical prosthesis.

TV prosthetics is associated with high mortality, especially in children under 1 year of age [25]. A significant disadvantage of prosthetics with biological valves in children is calcification, especially during the period of active growth and increase in the level of hormones in the blood. The need for continuous use of anticoagulants in case of implantation of a mechanical prosthesis gives serious complications and deterioration in the quality of life of young patients [26]. In addition, with the growth of the child, a discrepancy between the size of the prosthesis and the size of the chambers of the heart occurs, which again requires re-prosthetics with implantation of a larger prosthesis; the use of prostheses in newborns is impossible. According to Russian data of Yu.N. Gorbatyh et al., the need for prosthetics in a 10-year period reaches 45–60% [27].

TV prosthetics is justified only in cases of inefficiency or inability to perform plastic surgery; therefore, there was a great need for new valve plastic methods.

K. Hardy can be considered a pioneer in valve reconstruction with EA, who in 1964 first put forward the concept of reconstructing the valve from its own reduced tissue of the cusps, and also proposed to exclude the atrialized part of the ventricle [28]. In our country, the first attempt to perform valve plastic surgery was made by E.N. Meshalkin, using a strip of Ayvalon to restore the posterior and septal cusps, but the result was unsatisfactory, and only in 1978 I.K. Okhotkin reconstructed the valve. In 1979, based on the concept of K. Hardy, a team of surgeons from the Mayo Clinic (USA) under the direction of Professor G. Danielson developed the most advanced technique for that time.

The original method consisted of vertical plication of the atrialized part of the RV, "pulling" of the functional fibrous ring to the true fibrous ring using U-shaped sutures on pads, and narrowing of the fibrous ring along the anteroposterior commissure (Fig. 4). This technique is applicable provided there is sufficient mobility of the anterior tricuspid valve cusp (leaflet). As a result, a single-leaflet valve is formed [29]. M. Brown et al. applied the G. Danielson method in 182 patients.

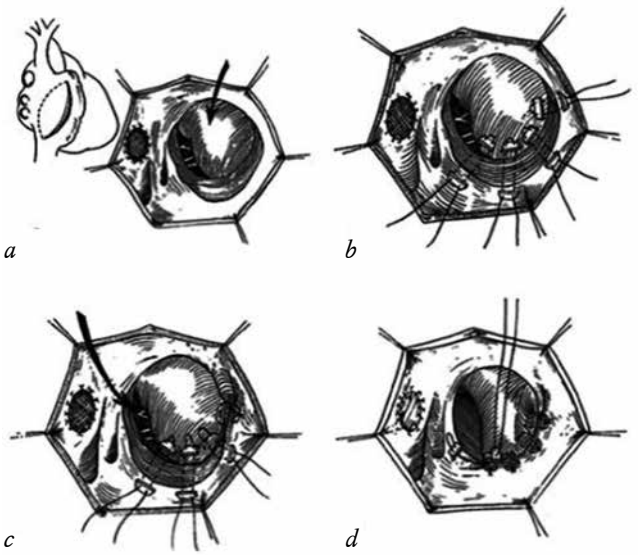

Fig. 4. Surgical correction according to the method of G. Danielson (*a–d*). Image modified from [29]

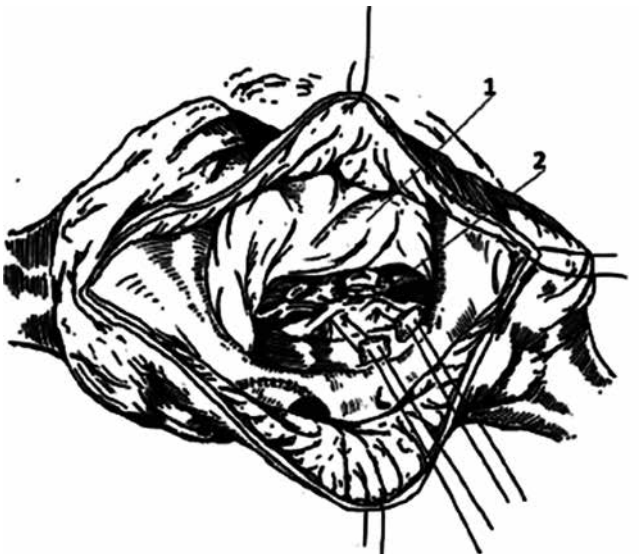

Fig. 5. Surgical correction according to the method F. Sebening stitch. Image modified from [31]: *1* – anterior cusp; *2* – plication of the posterior cusp

Mortality in the early postoperative period was 5%, over the course of 10 years – 12%. Over the next 20-year observation period, the number of reoperations with this method is 36%. This operation for a long time had no analogues for the correction of different types of EA.

The following technique was developed by the German surgeon F. Sebening. The essence of this method is to move the papillary muscle of the anterior cusp of the TV closer to the interventricular septum towards the true tricuspid ring (Fig. 5). The ultimate goal of this method is to create a single-leaflet valve. In this case, the anterior cusp, which is usually mobile, approaches the edge of the true fibrous ring [30].

This method can be combined with other methods of creating a competent TV, and it can be used as an independent method [31].

Takeushi Komoda (2007) used a combination of F. Sebening stitch and Hetzer techniques (see below). The study included 28 patients. In a group of 11 people, a combination of methods was used. Postoperative observation was carried out for 32 months. As a result, there was no postoperative mortality in the group with a combination of methods, and there were also no reoperations [32, 33].

The techniques of G. Danielson and F. Sebening stitch were similar and consisted of creating a valve by plicating the cusp tissue from the ventricle. They had positive aspects, but led to deformation in the area of the interventricular groove, which could lead to compression of the right coronary artery and its branches. This method is not widespread, since it was applicable only in patients with types A and B, when the tissue of the posterior and septal cusps is sufficiently mobile, and the anterior cusp should be of sufficient area and not limited in the movement of the chords.

In 1988, the French heart surgeon A. Carpentier for the first time drew attention to the possibility of creating a valve using its own hypoplastic cusp tissue. In his innovative technique (Fig. 6), he first described the delamination of the anterior and posterior cusps of the TV by dissecting fibrosed chords from the walls of the myocardium, starting from the highest point (functional ring).

Dissection of fibrosed chords gives mobility to cusp tissue for subsequent distribution over the entire surface of the true fibrous ring. However, chords attached to the papillary muscle must remain intact in order to avoid cusp prolapse. Then, the atrialized part is plicated along the posterior wall of the RV and narrowing of the true fibrous ring of the valve. Previously delaminated cusps are sutured onto a true fibrous ring with a clockwise rotation. Thus, a bicuspid valve is created at the level of the true fibrous ring. In adult patients, the fibrous ring is additionally fixed with a stented support ring. S. Chauvaud et al. reported 9% mortality in the early postoperative period and 13% mortality during the 10-year period. The number of reoperations was 11% over the next 20 years [14].

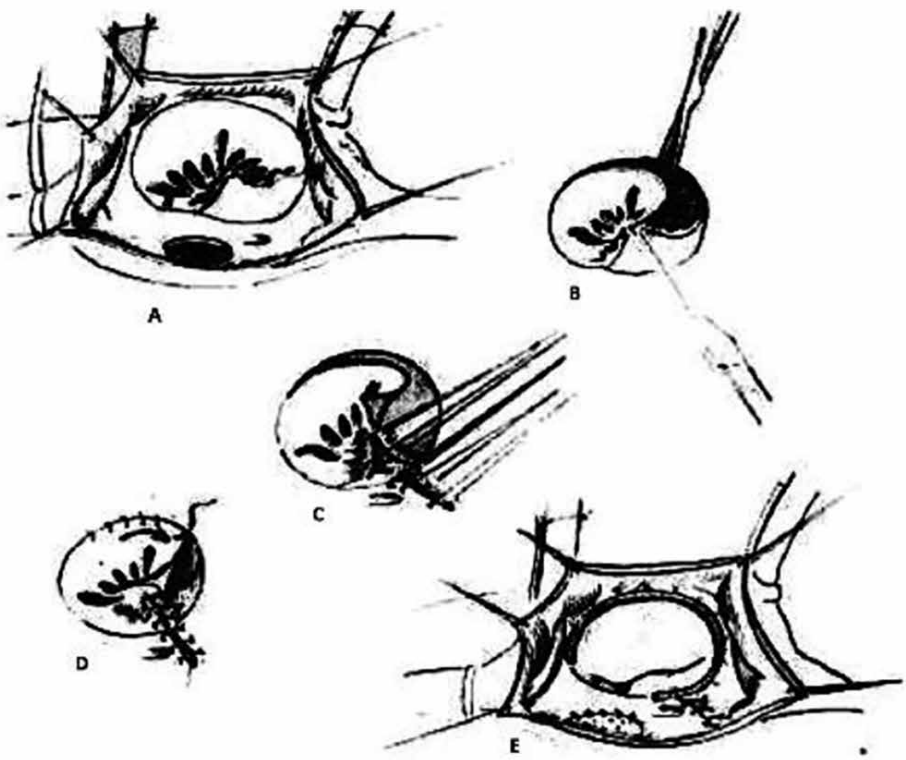

Fig. 6. Surgical correction according to the method of A. Carpentier (*a–e*). Image modified from [14]

The next intervention option was the technique of the German cardiac surgeon R. Hetzer, developed in 1998. The author describes various options for plastic surgery in his works, but the key point of his technique is suture plication of the TV. In this case, the posterior part of the TV is stitched with the septal part. In some cases, R. Hetzer suggested creating a valve of the double orifice type (Fig. 7). In this case, the cusp tissues are not mobilized to create a valve, and the atrialized part is not plicated; failure on the valve is reduced by reducing the surface area of the true fibrous ring [34]. The treatment results were published in 2015: mortality was 2.4% in the early postoperative period and 8.7% during the 10-year period. The number of reoperations left 7.1% over the next 20-year follow-up period [35]. In his works, R. Hetzer described various options for narrowing the fibrous ring, but the method of stitching the anterior and posterior parts of the fibrous ring is taken as a basis.

In 2000, the C.J. Knott-Craig method was published, which is used in the neonatal period by performing two-ventricular correction. C.J. Knott-Craig et al. performed surgical

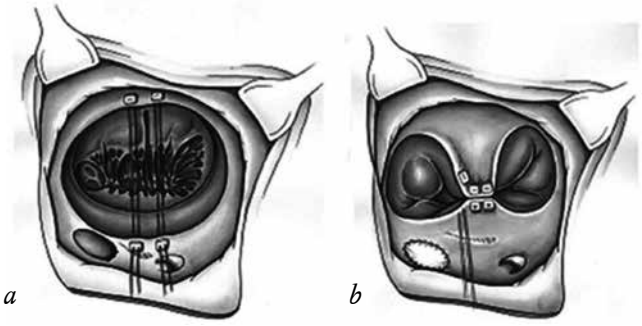

Fig. 7. Surgical technique of the double orifice (*a–b*). Image modified from [34]

treatment at an Oklahoma hospital on 27 children with EA, 22 of whom were newborns. The first step is the delamination of the front edge of the anterior valve cusp to ensure its mobility. Then, a suture is performed through the dominant papillary muscle of the anterior cusp with the bringing of the latter to the interventricular septum on the opposite side. To suture the enlarged fibrous ring, sutures are placed on the anteroposterior commissure of the valve or through the medial wall of the coronary sinus. This maneuver provides pulling the papillary muscle of the anterior cusp to the opposite

wall of the ventricle, and also gives greater freedom of the anterior cusp and increases its coaptation.

In the original method, a resection of the wall of the right atrium in the form of an ellipse is performed. When performing it in the lower corner of the incision, one should be careful not to damage the right coronary artery, since in newborns the border of the atrialized part of the ventricle and the right atrium is quite difficult to distinguish. Then, the atrialized part of the RV is sutured. The technique of TV plastic is similar to the technique of G. Danielson, the result of which should be the creation of a single-leaf valve. A prerequisite for this is the mobility of the anterior valve cusp [36].

If the anterior cusp has multiple chordal adhesions, then with an insufficient cusp length, it is possible to increase its surface using an autopericardial patch [37].

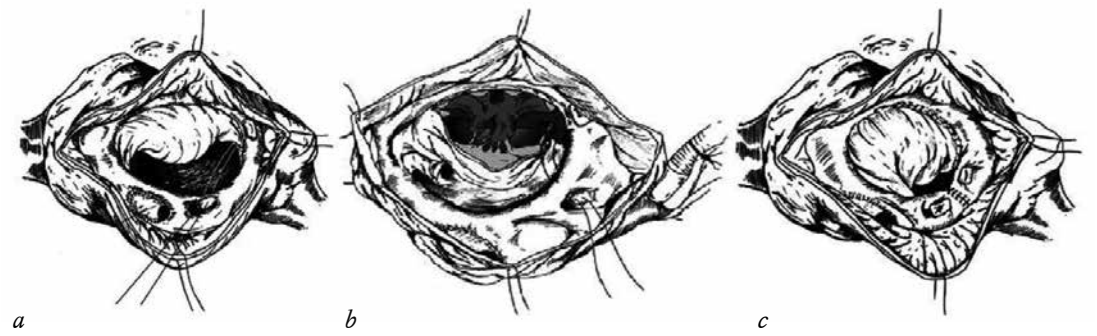

Fig. 8. Surgical correction according to C.J. Knott-Craig (*a–c*). Image modified from [36]

Atrial septal defect closure is usually performed with a defect of 4 mm left. In the neonatal period, the shunt has a discharge function for right-left discharge with right ventricular heart failure and increased pulmonary vascular resistance, which remains in newborns for the first few weeks of life.

In 2001, the arsenal of surgical interventions was supplemented by the technique of the Chinese cardiac surgeon Q. Wu. This technique was aimed at improving valve function due to the tissue of the posterior cusp. Q. Wu describes the technique as follows (Fig. 9): the posterior and septal cusps from the RV walls are mobilized, the cusps are stitched and attached to the level of the true fibrous ring, creating a bicuspid valve. In some cases, plastic on the septal cusp with an autopericardial patch was performed. In this technique, instead of plicating the atrialized part, a resection of the right ventricular triangular flap is performed [38, 39].

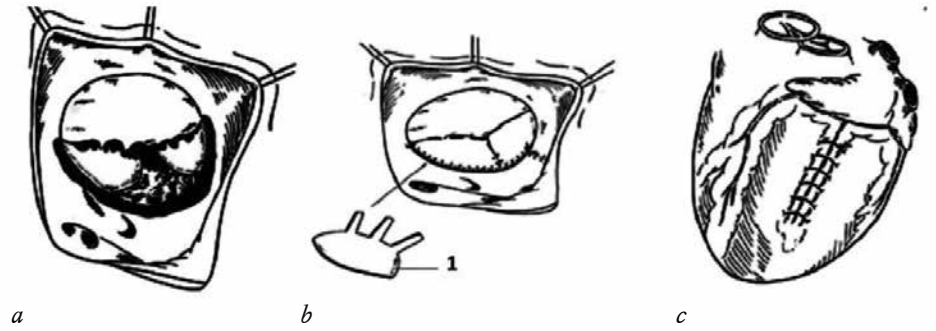

Fig. 9. Surgical correction according to the Wu method. Image modified from [38]: *a* – initial view of the valve (the delamination boundary is indicated by a dashed line); *b* is the final view of the valve; *c* – suturing of the atrialized part; *1* – autopericardial patch

According to the results of Dr. Q. Wu et al., the early and long-term postoperative period in 34 patients proceeded without complications. TV insufficiency was insignificant [40]. The use of this method has significant limitations, since in extreme forms the tissue of the septal and posterior cusps is practically absent. With resection of the atrialized part and suturing with a

twisted suture, the risk of postoperative bleeding and damage to the right coronary artery and its branches increases.

The most progressive method today is considered to be a "cone" reconstruction of the TV. This method was first proposed by the Brazilian cardiac surgeon J. da Silva in 1989. The basis for this reconstruction was the technique of A. Carpentier [41]. To date, the greatest experience in conducting "cone" reconstruction has been accumulated in the Mayo Clinic (Rochester, USA).

Surgical technique (Fig. 10) is as follows. Access to the TV is through the right atriotomy after connecting the heart-lung machine. Separation of the anterior cusp begins at 12.00 with the relative application of the clock-face to the TV, retreating from the fibrous ring a few millimeters. The incision continues clockwise towards the rear valve cusp. Fibrous and muscle adhesions between the anterior valve cusp and the right ventricle are dissected, releasing it. This is the most important surgical technique in the operation, since the maximum amount of tissue of the anterior cusp for subsequent plastic depends on its result. In some cases, longitudinal cuts are made 1/3–1/4 of the length from the edge of the anterior cusp, forming so-called neochords for better valve capacity in diastole.

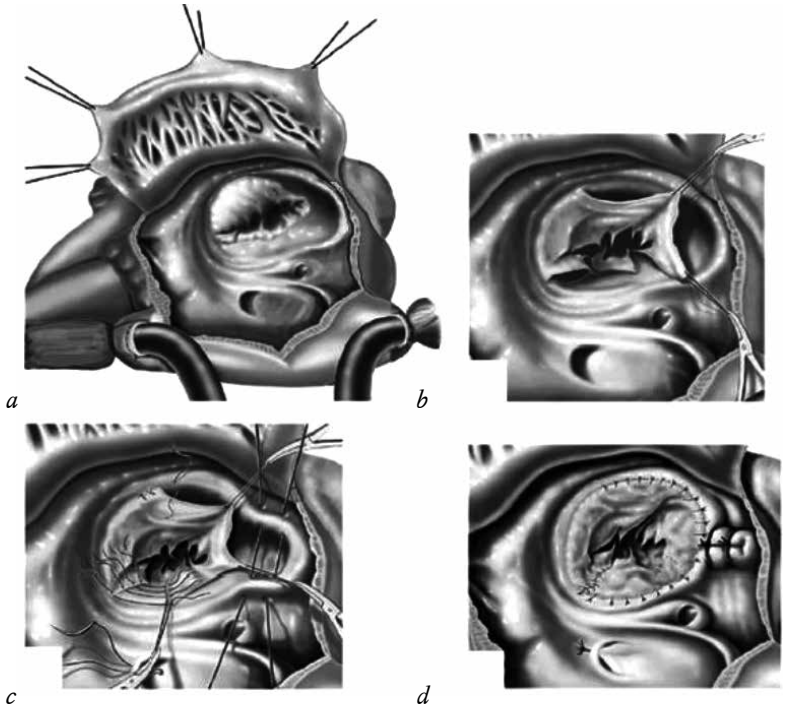

Fig. 10. Cone reconstruction of the tricuspid valve. Image modified from [42]: *a* – view of the valve before reconstruction; *b* – delamination of the anterior and posterior cusps; *c* – stitching of the valve cusps; *d* – final view of the valve in the form of a cone [41]

This technique is used when it is not possible to separate the edges of the cusp. After separation of the anterior cusp, the posterior and septal cusps follow, if possible. Upon completion of the process of delamination of the cusps, longitudinal plication of the atrialized part of the RV is performed with narrowing of the fibrous ring of the right atrioventricular valve. After plication, the mobilized tissue of the cusps is sutured together, forming a single cusp, which is distributed 360° around the circumference and is fixed to the true fibrous ring.

The cone reconstruction described by J. da Silva and colleagues differs from previous valvuloplasty methods in that it is closest to the "anatomical correction". The final result of the "cone" reconstruction includes a 360° distribution of TV tissue at the level of the true fibrous ring. This allows the TV cusps to close in the same way as in a normal valve. In addition, the reconstructed TV is attached to the true fibrous ring, so that the valve attachment point is now in its normal anatomical position. The thinned atrialized part of the RV is plicated, thus elimi-

nating the dyskinetic part of the RV. "Cone" reconstruction restores valve anatomy better than any method described above, and can be applied to a wide variety of valve anatomical variations encountered with Ebstein's anomaly [42].

The results of Dr. J. da Silva et al. described 52 patients operated on by this method. Early postoperative mortality was 3.8%, over the next 7 years – 14%, over the observation period - only 4 reoperations. C. Pizarro et al. reported about the implementation of the "cone" reconstruction on two children in the neonatal period, and one child was operated on before the age of 1 year. There was no postoperative mortality, and observation during the 1st year showed a positive trend according to echocardiography and chest x-ray. All patients retained functional capacity of the I class and TV deficiency under the 1st degree [43].

The article by K. Holst and J. Dearani (2018) presents the results of the treatment of 235 patients with "cone" reconstruction and a subsequent ten-year follow-up period. The study included 134 children and 101 patients over 18 years of age. In the early postoperative period there was one death (0.4%), the number of reoperations was 14 cases (5.9%). Long-term valve insufficiency was within the 1st – 2nd degree, statistically significantly decreased RV dysfunction and decreased apical diastolic RV area [44].

M. Ibrahim et al. on the example of 23 cases of "cone" reconstruction, they showed a decrease in insufficiency on TV and an increase in the end-diastolic volume of the left ventricle [45]. R. Lange et al. using echocardiography and magnetic resonance imaging showed an increase in stroke volume and a decrease in the size of the RV 6 months after this treatment method [46].

In Russia, this technique is actively used in cardiac surgery clinics in Tomsk [47], St. Petersburg [48], and Samara [49]. In the Cardiology Research Institute of Tomsk, "cone" reconstruction has been used since 2011; during this period more than 40 cases of its implementation have been accumulated.

With reduced RV function and inability to adequately provide pulmonary blood flow, a bidirectional cavopulmonary anastomosis is indicated. The method is performed as follows: the superior vena cava is cut off from the right atrium 0.5–1 cm above its entry (to exclude damage to the sinus node), the right atrium is sutured. The right pulmonary artery is dissected along, strictly above the superior vena cava, and sutured to the superior vena cava [50, 51]. The functions of the anastomosis of the superior vena cava and the right pulmonary artery are as follows: reduction in preload on the right ventricle (in childhood by about 1/2 venous return and 1/3 in adulthood); increased preload on the left ventricle.

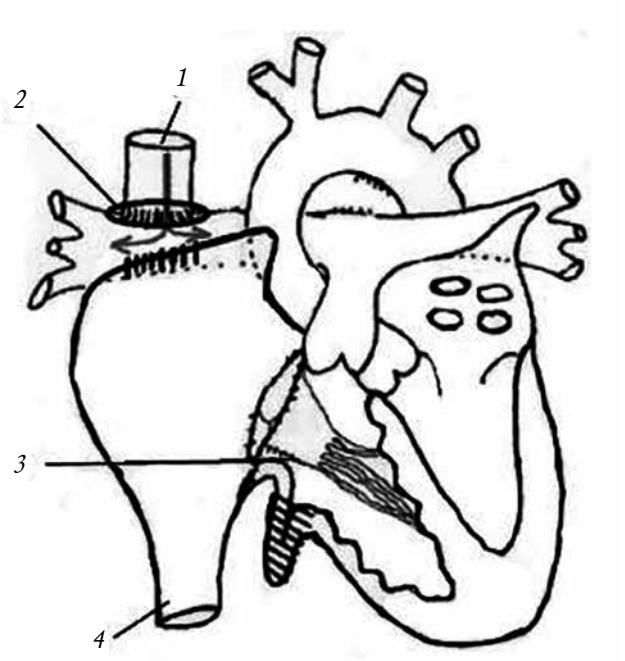

Fig. 11. Bidirectional cavopulmonary anastomosis in combination with the "cone" reconstruction of the tricuspid valve: *1* – superior vena cava; *2* – bidirectional cavapulmonary anastomosis; *3* – cone reconstruction of the tricuspid valve; *4* – inferior vena cava

CONCLUSION

Surgical treatment of Ebstein's anomaly has been a problem for more than half a century. After C. Bernard replaced TV with a prosthesis for the first time, many different TV techniques were developed. The first attempts at valve plastic surgery were made by S. Hunter and W. Lillehei in 1957. Valve prosthetics can be called a "desperate operation" when the surgeon is unable to construct a new valve from its own tissues. There are works on the use of own autopericardial tissue and the use of bone marrow cells - the precursors of $CD133^+$ endotheliocytes in the treatment of EA. In Russia, this technique is being developed at the A.N. Bakulev National Medical Research Center of Cardiovascular Surgery under the supervision of Academician L.A. Bokeriya [52].

The accumulation of experience and the improvement of surgical techniques have led to significant progress in the treatment of this heart disease. To date, according to world literature, the method of choosing the surgical treatment of EA is a "cone" reconstruction, which shows good results both in the near and in the distant postoperative period. However, debatable questions remain about the appropriate anatomy of the defect, the limiting possibilities of mobilizing cusp tissue for correction, and indications for performing a bidirectional cavopulmonary anastomosis. The ability to perform a "cone" reconstruction or other type of valve repair for Ebstein's anomaly also depends on the anatomy of the valve and subvalvular apparatus.

The most important feature for performing valve plastic surgery is the free, non-adhered edge of the anterior valve cusp, from which the largest volume of tissue is obtained. Reconstruction of the valve is difficult if the anterior cusp is strongly displaced to the apex of the RV, and there are extensive fibrous adhesions of the valve cusp with the adjacent myocardium. In his work, J. Stulak et al. indicate that the necessary conditions for successful correction are: delamination of the anterior cusp of more than 50% of its area; the presence of long chords and papillary muscle, which does not cause obstruction of the outflow tract of the right ventricle. The more tissue can be obtained by separating the cusp from the myocardium, the smaller the gradient and insufficiency on the valve can be expected in the end.

Thus, the "cone" reconstruction is considered the most "anatomical" of all existing (today) types of correction, and can be performed for a wide range of anatomical variants of Ebstein's anomaly in both childhood and adulthood.

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