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EBSTEIN'S ANOMALY IS A RARE CONGENITAL HEART DEFECT

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Absract. The lecture presents the history, epidemiology, etiology and pathogenesis, classification, clinic, methods of surgical treatment and rehabilitation of patients with Ebstein's anomaly — a rare congenital heart defect of the "blue" type, the incidence of which is 5.2 cases per 100 thousand newborns, which is about 1% of all congenital heart defects. The schemes of hemodynamics in this defect, the results of the examination, including visualization methods, the stages of surgical treatment are given.

Key words: *congenital heart defects; Ebstein's anomaly; children; tricuspid valve plastic surgery.*

АНОМАЛИЯ ЭБШТЕЙНА — РЕДКИЙ ВРОЖДЕННЫЙ ПОРОК СЕРДЦА

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Резюме. В лекции представлены история, эпидемиология, этиология и патогенез, классификация, клиника, методы хирургического лечения и реабилитации пациентов с аномалией Эбштейна — редкого врожденного порока сердца «синего» типа, частота встречаемости которого — 5,2 случая на 100 тыс. новорожденных, что составляет около 1% от всех врожденных пороков сердца. Приведены схемы гемодинамики при этом пороке, результаты обследования, в том числе визуализирующими методами, этапы оперативного лечения.

Ключевые слова: *аномалия Эбштейна; дети; пластика трикуспидального клапана; врожденные пороки сердца.*

Congenital heart diseases (CHD) are structural developmental anomalies of heart or great vessels that develop in an early intrauterine period [1]. CHDs are the second most common ones after congenital malformations of the nervous system [2]. About 17,500 children with various heart anomalies are born annually in the Russian Federation, which accounts for 249 in 100,000 infants. Congenital malformations are caused by exogenous factors in 89%, they include radiation, viral infections, maternal diseases during pregnancy, drugs and chemicals, and heavy metals. 10% of congenital heart diseases are due to inherited chromosomal abnormalities or may be the result of monogenic mutations. The diseases lead to heart dysfunction, blood stasis in veins, tissues and organs [3].

Ebstein malformation (EM) is a rare congenital cardiovascular defect, usually involving the tricuspid valve and right ventricle, with a wide range of anatomical and pathophysiologic manifestations [4]. This pathology is caused by incomplete delamination of the septal and posterior tricuspid valvar (TV) leaflets from the endocardium of the right ventricle (RV) at the stage of embryonic development. In turn, it leads to characteristic changes in

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Fig. 1. Wilhelm Ebstein, 1885 (© Städtisches Museum; Goettingen, Germany) [6].

Рис. 1. Вильгельм Эбштейн, ок. 1885 года (© Städtisches Museum; Геттинген, Германия) [6]

the valve apparatus: shortening and/or absence of TV chordae, apical displacement of the leaflets and coaptation zone, dilatation of the tricuspid valve. Moreover, EM reduces the functional part of the RV due to atrialization and progressive formation of myocardial fibrosis in the LV myocardium [5]. The incidence rate is 5.2 in 100,000 newborns, which is about 1% of all congenital heart defects. According to various authors, the average life expectancy in the natural course of the disease is up to 50 years, with 80–87% of fatal outcomes at the age of 30–40 years [6]. Since the pathology was first described by Wilhelm Ebstein (Fig. 1) in 1866, the malformation was named in his honor.

On June 28, 1864, a 19-year-old laborer was admitted to Dr. Ebstein's hospital, the patient had dyspnea and palpitations since childhood, which had increased with age. The patient's attending physician distinguished the patient's cachexia, marked cyanosis, jugular veins pulsing synchronously with the heart rate, a systolic heart murmur, and an enlarged cardiac silhouette detected during the percussion. The clinical picture indicated a congenital heart disease. Eight days later, the patient died. In 1866, Wilhelm Ebstein published his report, "A very rare case of tricuspid regurgitation caused by a congenital defect" [7].

ETIOLOGY

The exact causes of Ebstein's anomaly are unknown. The inheritability of Ebstein's anomaly is believed to obey Mendel's laws. The pathogenetic theory explaining the development of the desease is the concept of the abnormal cell death. There are observations that mothers using lithium gave birth to children with this anomaly. Lithium is known to be a toxic inducer leading to the birth of children with pathologic accessory conduction pathways. The use of benzodiazepines during pregnancy is another factor associated with a higher incidence of children born with this malformation. The risk is also higher in white families who have been exposed to paint products and have a history of miscarriage [8].

CLASSIFICATION

Currently, there are two main classifications widely used in cardiac surgical practice: anatomic and echocardiographic ones.

Troshkintsev, Podoksenov et al. cite Alain Carpentier's anatomic classification of Ebstein's malformation, according to which four types were distinguished (presented in Fig. 2).

The echocardiographic classification, for the first time proposed by Selermeier et al. is based on the correlation between the degree of the atrialized part of the LV and its functional part, as well as the LV in the four-chamber position at the end of diastole [9]. This correlation has four gradations depending on the degree of severity:

ratio 0.5 — I degree;
ratio 0.5 to 0.99 — II degree;
ratio from 1 to 1.49 — III degree;
ratio of more than 1.5 — IV degree.

Such gradation allows predicting the natural course of the malformation in patients: in case the ratio is more than 1.5, the probability of lethal outcome is 100%; the ratio from 1 to 1.4 corresponds with the lower probability of early lethal outcome and, as a rule, is up to 10%, however the mortality rate in the early childhood can reach 45%; in case the ratio is less than 1, the probability of lethal outcome is lower, the survival rate in patients reaches 92% [9].

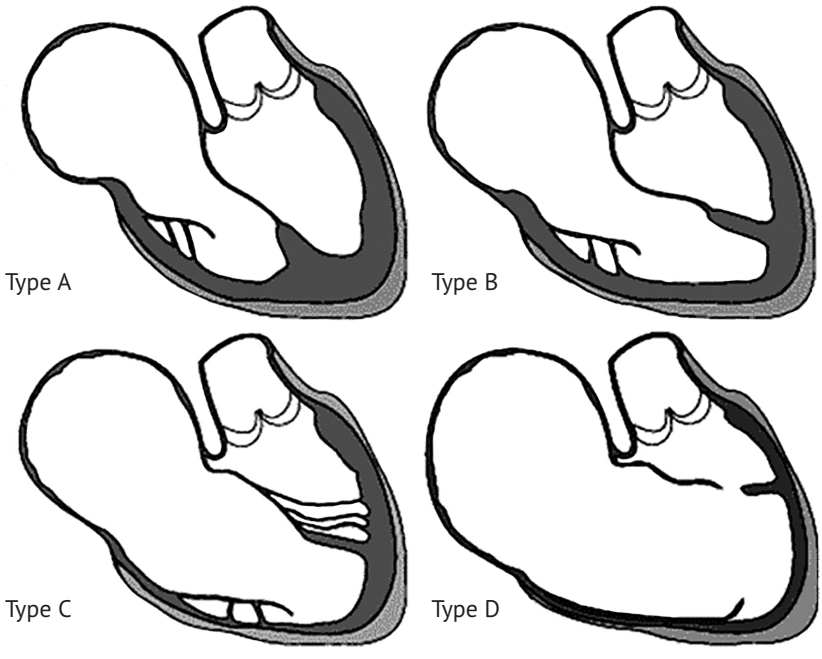

Fig. 2. Types of Ebstein malformation: type A — the volume of the true right ventricle (RV) is satisfactory; type B — there is a large atrialized component of the RV, and the anterior leaflet of the TV moves freely; type C — the anterior leaflet of the TV is limited in its movement and may cause obstruction in the RV outflow tract; type D — almost complete atrialization of the ventricle excepting a small part of the LV outflow tract [7]

Рис. 2. Типы аномалии Эбштейна: тип А — объем истинного ПЖ удовлетворительный; тип В — имеется большой атриализованный компонент ПЖ, а передняя створка ТК свободно перемещается; тип С — передняя створка ТК ограничена в своем движении и может служить причиной обструкции в выводном отделе ПЖ; тип D — почти полная атриализация желудочка за исключением небольшой части выводного отдела ПЖ [7]

PATHOLOGICAL ANATOMY

With respect to the norm, the tricuspid valve consists of three leaflets: anterior, posterior, and septal. Ebstein's malformation include following features (Fig. 3):

- There is insufficient detachment of the tricuspid valve leaflets which results in an anterior and apical rotational displacement of the functional valve ring. Downward displacement of the septal and posterior leaflets into the right ventricle is the main feature of this malformation.
- The anterior leaflet is usually attached to the TV but is enlarged or sail-like. In addition, the leaflet has multiple perforations and is fixed by chordae to the parietal wall of the ventricle.
- The part of the RV above the functional valve («atrialized right ventricle») is dilated, dysplastic, thin, with areas of hypertrophy. The tricuspid valve is almost always enlarged.
- The RV cavity behind the "atrialized" part is reduced ("functional right ventricle"), the inflow part is absent with preservation of the trabecular section.
- The right ventricular outflow tract is often obstructed by excess tissue of the anterior

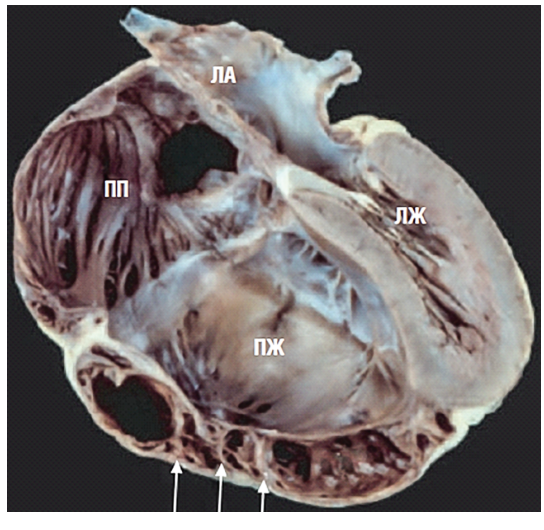

Fig. 3. Macropreparation of the heart in Ebstein's malformation. There is marked dystopia of the tricuspid valve leaflets with dilatation of the right ventricle at the expense of the atrialized part. LV — left ventricle; LA — left atrium; RV — right ventricle; RA — right atrium. The arrows show the obstruction of the right ventricular outflow tract [9].

Рис. 3. Макропрепарат сердца при аномалии Эбштейна. Отмечается выраженная дистопия створок трикуспидального клапана с дилатацией правого желудочка за счет атриализованной части. ЛЖ — левый желудочек; ЛП — левое предсердие; ПЖ — правый желудочек; ПП — правое предсердие. Стрелками изображена обструкция выводного отдела правого желудочка [9]

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leaflet and its chordal attachments, causing true anatomic obstruction. The obstruction is sometimes functional as a result of extremely poor contractility of the right ventricle [10].

PATHOPHYSIOLOGY

The degree of hemodynamic impairment in Ebstein's malformation is determined by the severity of anatomo-morphologic changes in the tricuspid valve and the RV. During systole of the right atrium, the atrialized chamber of the right ventricle is in the diastole phase. These discordant contractions lead to hindrance of blood flow in the atrialized chamber of the RV, causing decreased efficiency of atrial systole. Deformation of displaced tricuspid valve leaflets and dilated fibrous ring leads to its pronounced insufficiency. A large volume of venous blood during systole returns to the right atrium. Impeded outflow of blood from the right atrium leads to venous stasis in the great circulation circle and venous blood discharge into the left atrium through the open oval window or the atrial septal defect (ASD). The discharge of venous blood into the left heart causes arterial hypoxemia, which is directly proportional to pressure gradient between the atria. The blood flow into the left atrium through the ASD can be considered as a compensatory mechanism delaying the development of systemic venous insufficiency. The closer the tricuspid valvar leaflets are displaced to the apex of the RV and the smaller the diameter of the interatrial junction is, the more severe hemodynamic disorders develop in the patient [11].

CLINICAL PICTURE

Clinically, Ebstein's malformation can be manifested by dyspnea, skin cyanosis, pronounced cardiomegaly, decreased tolerance to physical exertion and the development of heart failure. However, the symptoms of the malformation might be absent and manifest themselves in a later period.

The clinical heterogeneity of patients with EM varies according to the severity of tricuspid valve pathology, the size of the functioning RV, the speed of the blood flow due to obstruction between the inflow and outflow tracts of the RV, the ammount of blood discharge at the atrial level, the presence of cardiac rhythm disturbances and concomitant heart defects [12, 13].

Clinical severity often correlates with the age when a patient is admitted to the hospital for the first time. Intrauterine, neonatal and infantile age are more often accompanied with additional diagnoses and more severe manifestations with cyanosis, right ventricular failure and high mortality. Dyspnea, fatigue, ascites, edema, exercise intolerance, and cyanosis might be present in children, adolescents and adults, which correlates with disease severity, but most of the symptoms attract attention when atrial arrhythmias appear [14]. Interatrial communication significantly increases the risks of paradoxical embolism, brain abscess and sudden death along with decreased exercise tolerance, which is determined by the volume of the intracardiac shunt and the degree of blood oxygenation in the systemic blood flow [9]. Increasingly, deformation of the terminal phalanges of fingers in the form of "drumsticks" (in children older than 1 year) is noted.

Jugular vein pulsation during examination which occurs as a result of marked regurgitation of the tricuspid valve and large volume overload of the right atrium is rarely observed. The third and fourth cardiac tones are regularly present in the act of auscultation [15–17]. Additionally, the first and second heart tones are often split due to delayed tricuspid and pulmonary components. Patients with EM often have a holosystolic murmur [15–17]. Thickening of the phalanges of the fingers in Ebstein's malformation depends on cyanosis and is caused by the degree of hypoxia.

ELECTROCARDIOGRAPHY

Supraventricular tachycardia (SVT) is the most frequent rhythm disturbance. It occurs in 7–30% of patients. This type of arrhythmia is due to the presence of additional conduction pathways (ACP) or additional atrial-ventricular junctions (AVJs) such as Wolff-Parkinson-White (WPW) syndrome. Other various types of tachycardia are also found in patients with the Ebstein's malformation: atrial ectopic tachycardia (AET), atrial flutter (AF), atrial reentry tachycardia (ART), atrial fibrillation and ventricular tachycardia [18]. Complete atrioventricular heart block is rare in the Ebstein's malformation, but I degree atrioventricular block is reported in 42% of patients due to dilatation of the right atrium [19, 20]. The atrioventricular node in the Ebstein's malformation may be prone to compression due to abnormal formation of the central fibrous body. Anomalies of the right bundle branch of Hiss and fibrosis of this part of the conducting system may be noted [21–23]. Between 6 and 36% of patients with Ebstein's malformation have at least one accessory conduction pathway [24–28], most

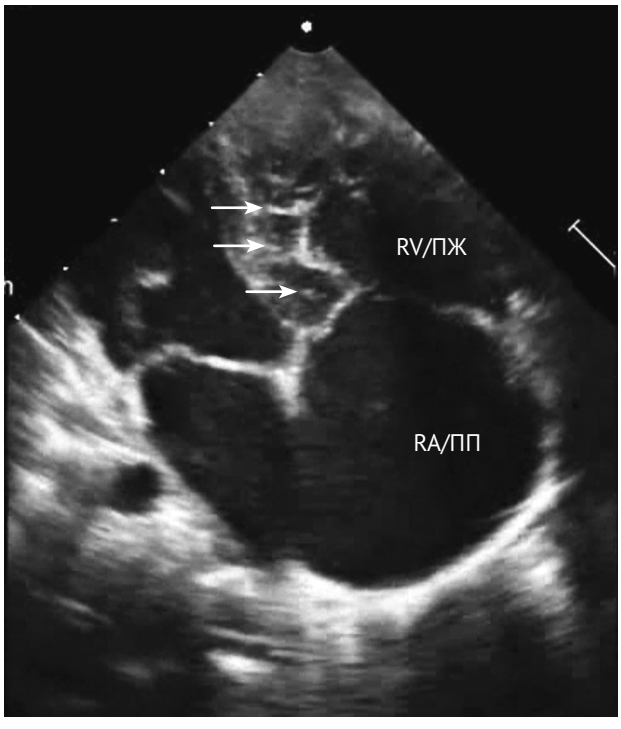

Fig. 4. Displacement of the right atrioventricular ring into the right ventricle (RV). Dilated RV and right atrium (RA). Delamination disorder of the septal leaflet of the TV (arrows)

Рис. 4. Смещение правого атриовентрикулярного кольца в правый желудочек (ПЖ). Дилатированный ПЖ и правое предсердие (ПП). Нарушение деламинации септальной створки ТК (стрелки)

of the accessory pathways are located around the open atrioventricular opening of the tricuspid valve [29–31]. Tachycardia with a wide QRS complex is due to supraventricular and septal accessory conduction pathways, which may cause ventricular tachycardia and flutter, ectopic atrial tachycardia, and atrial fibrillation [26, 27].

ECHOCARDIOGRAPHY

Diagnostic criteria:

- one or more TV leaflets are displaced to the apex of the RV;
- The point of closure of the TV leaflets is displaced to the apex by more than 8 mm;
- Signs of atrialization of the LV;
- Lengthening of one or more TV leaflets;
- TV dysfunction;
- Atrial septal defect (ASD) [32].

When pulmonary artery and aortic valve motion are simultaneously reordered, aortic valve closure occurs significantly earlier than pulmonary artery valve closes. Late pulmonary valve closure can be explained by reduced pumping function of the right ventricle due to its reduced size, high residual diastolic pressure and low compliance.

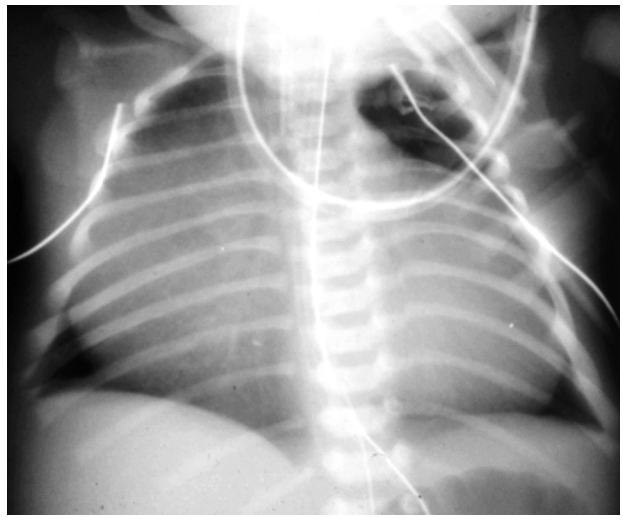

Fig. 5. Preoperative chest X-ray of a patient with Ebstein's malformation. A globular configuration of the heart with a pronounced expansion of borders in the cross-section is observed

Рис. 5. Обзорная рентгенография грудной клетки пациента с аномалией Эбштейна до операции. Отмечается шаровидная конфигурация сердца с выраженным расширением границ в поперечнике

The interatrial septum may displace into the left atrial cavity. The functional part of the right ventricle may compensatory enlarge and compress the left ventricle with further obstruction of the outflow tract.

2D-echocardiography is the most informative. Displacement of the right atrioventricular annulus is well visualized in a four-chamber view. The displacement can have varying degrees. With minimal displacement, the Ebstein's malformation is detected incidentally because there are no significant hemodynamic abnormalities [33] (Fig. 4).

RADIOLOGIC EXAMINATION

The cardiac shadow may vary from a nearly normal configuration to the globular cardiac configuration typical for Ebstein's malformation. There are signs of right atrial enlargement (upward shift of the right cardiovasal angle) [9]. Narrow vascular plexus and depression of the pulmonary artery segment. The left heart sections are not enlarged. Blood supply of the pulmonary fields may be normal or reduced. When cardiothoracic index is more than 65% it is considered as a prognostically unfavorable sign. There may be marked cardiomegaly (a box heart) with the heart silhouette almost filling the entire chest [10] (Fig. 5).

CARDIAC CAVITY CATHETERIZATION

Many patients with Ebstein's malformation may develop complex cardiac rhythm abnorma-

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lities during invasive cardiac testing that can result in cardiac arrest [34]. Diagnostic cardiac cavity catheterization is rarely required in patients with Ebstein's malformation and is mainly performed preoperatively for the purpose of coronary angiography. The pressure in the right ventricular and pulmonary artery in patients with Ebstein's malformation is usually normal, the right ventricle may have specifically increased end-diastolic pressure. Right atrial pressure may be normal despite severe regurgitation at the tricuspid valve, especially when the right atrium is discernibly dilated. Decreased systemic arterial oxygenation may be noted in the presence of interatrial communication and a marked right-to-left blood shunt during oxyhemometry.

TREATMENT

Peculiarities of drug therapy

Patients with Ebstein's malformation and heart failure who do not require surgical treatment are treated with standard therapeutic tactics for heart failure. The efficacy of angiotensin-converting enzyme (ACE) inhibitors in patients with Ebstein's malformation who have right ventricular heart failure has not been proven. Drug treatment of arrhythmias should be individualized and combined with surgical or endovascular methods of treatment when indicated [9].

Peculiarities of surgical treatment

Indications for surgery concerning EM are:

- the presence of a clinical picture in the neonatal period;
- at older age, surgical treatment is indicated in case of:
  - decreased tolerance to exercise;
  - right ventricular heart failure (NYHA class III–IV);
  - cyanosis on exercise;
  - arrhythmias uncontrolled by drug therapy occur;
  - decreased LV function due to its compression by the interventricular septum since the LV is dilatated [32].

The main goal of surgical intervention is to restore the closing function of the tricuspid valve, as well as to increase the volume of the RV, reduce the volume of the right atrium cavity and close the interatrial septum.

Treatment of severe Ebstein's malformation in newborns includes the use of prostaglandin E to prevent closure of the ductus arteriosus and administration of vasodilators. In case of insufficient interatrial communication, endovascular dilatation of the open oval window or the atrial septal defect is indicated (Rashkind procedure).

Relative contraindications for correction: age over 50 years; severe pulmonary hypertension; significant decrease in left ventricular function (ejection fraction less than 30%); complete impairment of delamination of septal and posterior tricuspid valve leaflets, with less than 50% delamination of the anterior leaflet [35].

There are two concepts for surgical treatment of Ebstein's malformation:

- univentricular correction;
- biventricular correction.

Single ventricle correction

The aim is to exclude the right ventricle from the bloodstream with further Fontaine surgery. Indication for univentricular correction of the defect is a severe clinical manifestation of Ebstein's malformation in a newborn, when the prognosis for life during the first month without surgery is unfavorable.

It is considered that main advantages of including the hypoplastic (pulmonary) ventricle for partial support of pulmonary circulation are [36]:

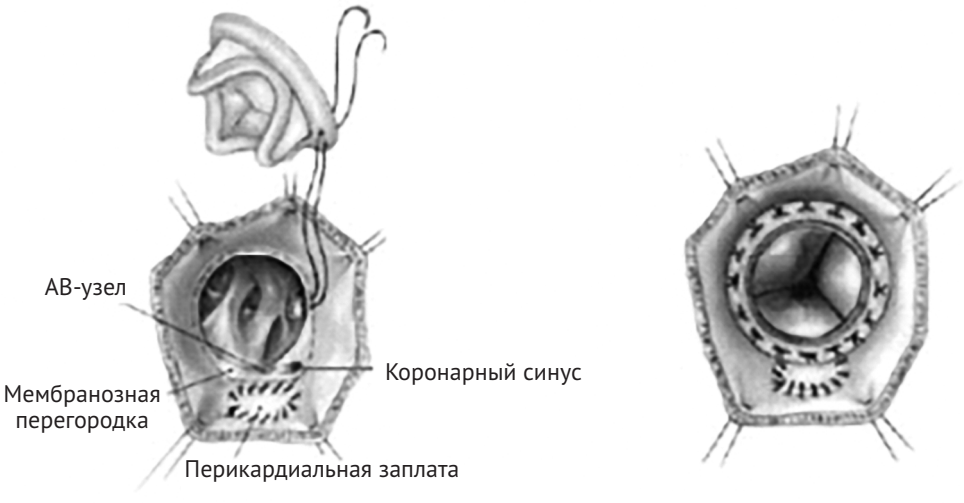
1) the possibility to increase cardiac output;
2) adaptation to exercise;
3) maintenance of pulsatile flow in the pulmonary circulation.

Patients who have safely survived the neonatal period need dynamic monitoring in the absence of surgical treatment indications [32].

Biventricular correction (cone reconstruction)

In most cases of biventricular correction, operative treatment consists of:

1) tricuspid valve repair;
2) plication of the atrialized part of the LV;
3) the right atrium repair [32];
4) the atrial septal defect repair or open oval window (OOW) repair (in case of moderate RV insufficiency with the fistula remaining about 3 mm);
5) elimination of previously performed systemic-pulmonary shunts and correction of concomitant anomalies such as ventricular septal defect (VSD), stenosis or open aortic duct (OAD) in accordance with the principles of their isolated correction;
6) performing various antiarrhythmic procedures when indicated (radiofrequency ablation or surgical dissection of accessory conduction



a / а *b / б*

Fig. 6. Schematic representation of bioprosthetic tricuspid valve replacement in Ebstein's malformation. Along with the implantation of the bioprosthesis, the atrial septal defect is closed with a pericardial patch: *a* — the process of tricuspid valve replacement; *b* — the result of tricuspid valve replacement [9]

Рис. 6. Схематическое изображение биопротезирования трикуспидального клапана при аномалии Эбштейна. Наряду с имплантацией биопротеза осуществляют закрытие дефекта межпредсердной перегородки перикардиальной заплатой: *а* — процесс биопротезирования трикуспидального клапана; *б* — результат биопротезирования трикуспидального клапана [9]

pathways, cryoablation of the atrioventricular junction, right-sided MAZE procedure).

Biventricular correction of Ebstein's malformation always raise the issue of preserving the native valve and performing reconstructive surgery or prosthetic valve replacement. Some authors report the possibility of plastic surgery in virtually any anatomy of the malformation, while others recommend replacing the native valve with an artificial one if there is the slightest doubt, moreover the latter have not completely managed the issue of choosing an artificial valve, although most cardiac surgeons prefer biological prostheses.

Valve replacement is possible with the use of biological or mechanical prosthesis (Fig. 6) [37]. The prosthetic valve replacement is specific since the valve is located above the true fibrous ring of the TV. The tissue of leaflets causing obstruction of the RV outflow tract should be necessarily excised, and the true fibrous ring should be narrowed to the size of the prosthesis. The atrialized portion of the RV is reduced as well. The posterolateral wall tissue is usually thinner, so the suture line should be closer to the atrium to avoid injury to the right coronary artery. In order to avoid damage of the atrioventricular plexus, the suture line is placed above the coronary sinus, so that venous blood drainage will be in the RV.

Bioprostheses should be favored in prosthetic issues, but the use of mechanical prostheses is also justified in patients taking anticoagulants for unrelated reasons [38]. The technique of TV replacement currently performed is presented in Fig. 6.

However, most authors agree on the preference for reconstructive interventions on the TV [37, 39, 40], as they demonstrate a more durable and physiologic result and avoid potential complications inherent to valve replacement.

The surgical technique is carried out as follows. The operation is performed from the midline access. Access to the TV is performed through the right atriotomy after connection of the artificial circulation apparatus. The anterior leaflet of the TV is cut off from the true fibrous ring of the TV in the zone of «10 o'clock», the incision continues clockwise to the posterior leaflet. The leaflets are detached from the right ventricular myocardium by sharp and blunt routes, all secondary chordae fixing the leaflets are crossed, and leaflet delamination is performed. In case the anterior leaflet edge is fused with the right ventricular myocardium, fenestrating incisions are performed to allow blood flow from the RA into the RV. Delamination of the septal leaflet is performed. The holes in the leaflets are sutured, the edges of the formed flaps are sutured. The cut off edge of the posterior leaflet is rotated clockwise by 180° and fixed to the edge of the septal leaflet. The reconstructed tricuspid valve has a cone shape. Excision or plication of the atrialized part of the right ventricle is performed, as well as plication of the TV fibrous

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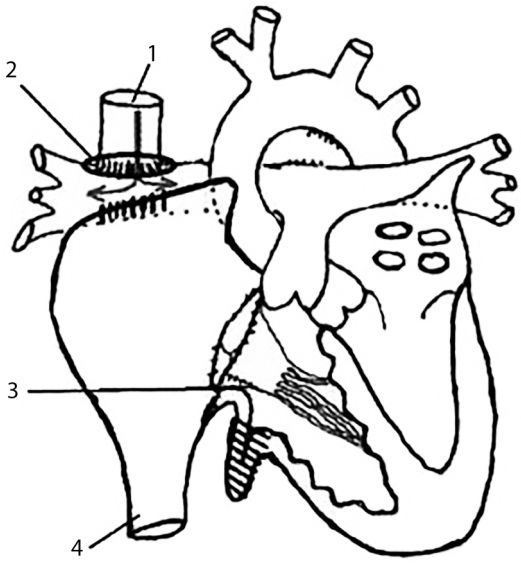

Fig. 7. Bidirectional cavapulmonary anastomosis combined with " cone" reconstruction of the tricuspid valve: 1 — superior vena cava; 2 — bidirectional cavapulmonary anastomosis; 3 — cone reconstruction of the tricuspid valve; 4 — inferior vena cava [7]

Рис. 7. Двунаправленный кавапульмональный анастомоз в сочетании с «конусной» реконструкцией трикуспидального клапана: 1 — верхняя полая вена; 2 — двунаправленный кавапульмональный анастомоз; 3 — конусная реконструкция трикуспидального клапана; 4 — нижняя полая вена [7]

ring is carried out, after which the cone is fixed to the true TV fibrous ring with a continuous single-row curling suture. In order to prevent atrioventricular conduction disturbance, sutures in the area of the membranous part of the interventricular septum are shifted towards the right atrium and superficially applied. If necessary, resection and plication of the RA are performed [41].

"Cone" reconstruction makes it possible to restore the valve closure function and can be applied to a wide variety of anatomical variants of pathology encountered in Ebstein's malformation [42]. In Russia, this technique is actively used in cardiac surgery clinics in Tomsk, St. Petersburg, and Samara.

Bidirectional cavapulmonary anastomosis is indicated in case of reduced LV function and inability to adequately provide pulmonary blood flow. The method is performed as follows: the superior vena cava is cut off from the right atrium 0.5–1 cm above its orifice (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 with the severed superior vena cava [43, 44]. The main objectives of this operation are to decrease the right ventricular preload (in childhood about 1/2 of venous return and 1/3 in adulthood) and to increase the left ventricular preload (Fig. 7).

ELECTROCARDIOSTIMULATION

BAbout 3.7% of patients with Ebstein's malformation require permanent electrocardiostimulation. The most common indication for pacemaker implantation is a complete form of atrioventricular block, and less frequently it is sinus node weakness syndrome [45]. In cases of artificial tricuspid valve replacement, the most expedient is to carry the electrode outside the cuff of the prosthesis. In cases of bioprosthetic tricuspid valve replacement, the electrode can be placed through the valve orifice, but this method is undesirable as the electrode may interfere with the normal mobility of the bioprosthetic leaflets and cause regurgitation.

TREATMENT OF ARRHYTHMIAS

Accessory conduction pathways or accessory atrial-ventricular junctions are noted in approximately 50% of cases [46–48] in EM patients. In addition, atrial flutter or fibrillation are frequent findings [49]. In the presence of premature ventricular excitation, there is a high risk of life-threatening ventricular arrhythmias. Prior tricuspid valve surgery may complicate catheter ablation of accessory pathways. Thus, even if preexcitation on ECG and history of supraventricular tachycardia are absent, it is reasonable to perform electrophysiologic study (EPS) before surgery to correct Ebstein's malformation and, if necessary, to perform catheter ablation before tricuspid valve surgery [50–52]. Although 3D mapping [53] and other advances in catheter technology are improving outcomes [53], recurrence rates remain high [54].

POSTOPERATIVE FOLLOW-UP

1. The duration and frequency of follow-up of patients with corrected Ebstein's malformation is determined individually. Hemodynamic status and the presence of arrhythmias should be evaluated regularly.

2. Prevention of bacterial endocarditis is necessary when indicated.

3. In the absence of signs of heart failure, it is acceptable to engage patients in physical training and sports after correction of the malformation [32].

FUTURE PROSPECTS

In most patients, right ventricular function improves postoperatively after an initial decline, regardless of the surgical strategy [55, 56]. In addition,

there is a tendency for gradual reduction of RV volume [55]. However, even early tricuspid valve repair fails to restore normal RV function in some cases.

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