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A Clinical Case of Successful Surgical Correction of Tetralogy of Fallot by Using the Right Atrial Appendage as a Neopulmonary Valve

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Abstract

Tetralogy of Fallot (TOF) is a common cardiac malformation characterized by reduced pulmonary circulation. Long-term preservation of neopulmonary bicuspid valve function is crucial in paediatric cardiac surgery. In 2019, Iranian cardiac surgeon Amirghofran A. introduced a novel method of forming a neopulmonary valve from the right atrial appendage. Despite numerous previous attempts to preserve the pulmonary valve using both autologous and foreign tissue, results were often disappointing. The usage of the right atrial appendage as a bicuspid valve in the pulmonary position offers an alternative method for correcting TOF with a deformed, hypoplastic, or aplastic pulmonary valve. However, further follow-up is required to obtain more reliable data.

Keywords: Tetralogy of Fallot, neopulmonary valve, right atrium appendage, right ventricular outflow tract.

Introduction

Tetralogy of Fallot (TOF) is a common cardiac malformation characterized by reduced pulmonary circulation. For many years, the surgical technique for correcting TOF remained unchanged. Despite numerous attempts to improve the technique, many proposed methods were not practical. In 2019, Iranian cardiac surgeon Amirghofran A. introduced a novel method of forming a neopulmonary valve from the right atrial appendage. This method serves as an alternative option for hypoplasia and aplasia of the pulmonary valve annulus. One notable benefit is the use of autologous valves, which may facilitate growth and potentially reduce the need for repeat interventions in children with pulmonary valve insufficiency and restenosis. [3-5]. In our opinion, one of the most promising techniques is the use of the right atrium appendage as a neopulmonary valve. More than 100 operations using this technique have been performed with good results, but it has not been used in post-Soviet countries.

Clinical case

The first successful surgical correction of Tetralogy of Fallot by using the appendage of the right atrium as a neopulmonary valve was performed in the paediatric cardiac surgery department of JSC NSMC (November 22, 2023) for the first time in Kazakhstan. Patient A., aged 11 months, was admitted with complaints of dyspnoea, sweating when feeding and crying, and lividity of the nasolabial triangle when restless. The examination according to echocardiography revealed the following: CHD. Tetralogy of Fallot: subaortic defect of the interventricular septum. Moderate tricuspid regurgitation. Aortic dextroposition of 50%. Combined pulmonary artery stenosis with trunk and branch hypoplasia. Gradient on the RV/PA is 85/42 mmHg. Condition after endovascular stenting of the RVOT. Hypertrophy of right ventricle myocardium. Ejection fraction (EF) is 75%. Left ventricular hypertrophy is present. Cardiac computed tomography angiography (CTA) with contrast reveals a functioning stent of the right ventricular outflow tract (RVOT), aortic dextraposition up to 50%,

RVOT obstruction, and myocardial hypertrophy of the right ventricle. Dilatation of all heart chambers is also observed. CTA findings are unremarkable, with no evidence of coronary artery obstruction, stenosis, or anomalies.

The duration of the operation was five hours, with a total time of artificial circulation of 178 minutes. The aortic clamping time was 78 minutes, with a blood loss of 15 ml. During the operation, the hypoplastic pulmonary artery trunk was opened and placed on holders. At the time of the revision, the pulmonary valve was bicuspid, the pulmonary leaflets were fused with the stent, and the leaflets were thickened and restricted in movement. Gradually, the valve leaflets were detached from the stent. Further, during revision through the pulmonary valve annulus, the bougie №5 was passed, which did not correspond to the calculated z-score parameters (z score parameters should be within 8-10 mm). It was therefore decided to cut the annulus and implant a neopulmonary valve formed from the appendage of the right atrium.

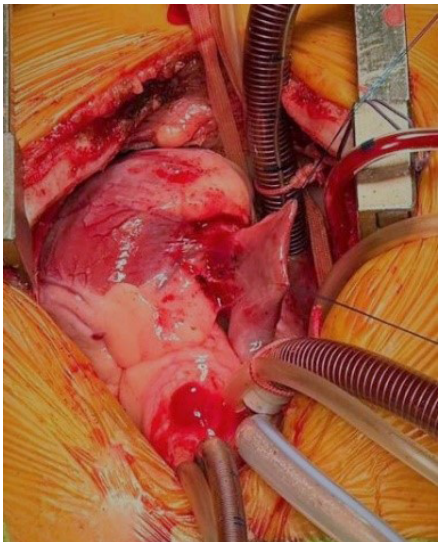

Figure 1 – Harvesting and preparation of the right atrial appendage (photo was taken during the operation)

Following the closure of the ventricular septal defect with a Gore-Tex patch and the excision of the right ventricular outflow tract by cutting hypertrophied and fibrotic tissues of the RVOT, the rudimentary pulmonary valve leaflets were excised, and a neopulmonary valve derived from the right atrium appendage was implanted in the location of the native pulmonary valve annulus (Fig. 1, 2, 3). The right atrium appendage was prepared in advance: The edges of the right atrium appendage were secured in position using prolene 5/0-10 thread. The rough portion of the right atrial appendage was then detached from the trabecular muscles. On the opposite edge of the appendage, a scalpel incision was made to correspond to the estimated diameter of the pulmonary valve annulus. Subsequently, the lateral edges of the neobicuspid valve were sutured in the projection of the pulmonary artery trunk. The next step involved suturing the lower edge of the leaflet of the neobicuspid valve to the pulmonary valve annulus. Subsequently, dilating plasty of the pulmonary artery trunk and right ventricular outflow tract was performed using xenopericardium. Additionally, the second leaflet of the neopulmonary valve was sutured to the xenopericardium in the region of the pulmonary valve annulus to preserve the haemodynamic geometry of the valve, thus avoiding neobicuspid valve deformation and insufficiency. On the control transoesophageal echocardiogram, the gradient on the RV/PA

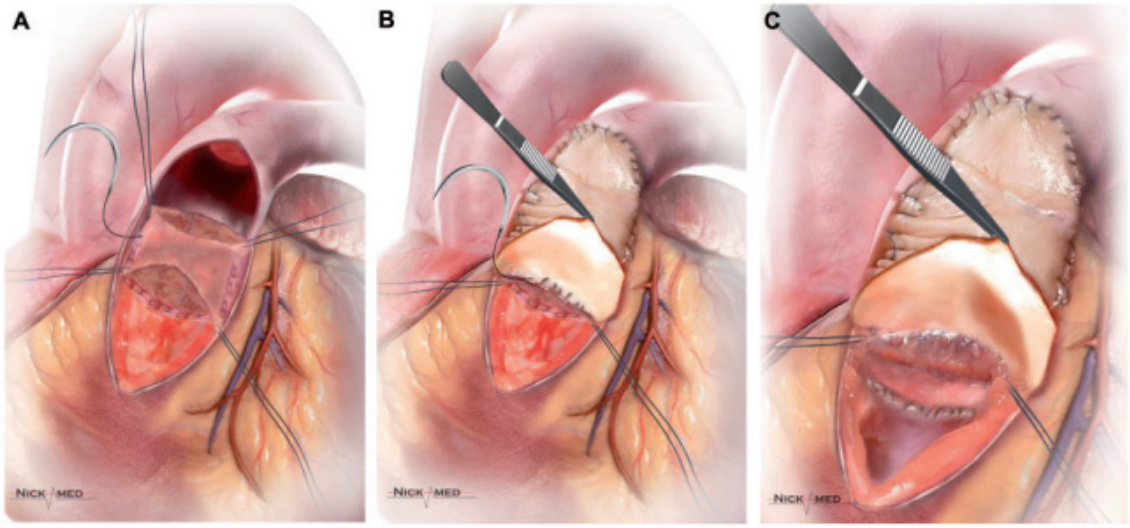

Figure 2 – Suturing the right atrial appendage valve in the place. (A) The posterior part of the annulus and then the two lateral edges are sutured to make the left and right commissures. (B) The bovine transannular patch covers the pulmonary arteriotomy, and the anterior annulus is sutured. (C) The final shape of the bicuspid right atrial appendage valve (the figure was taken from [1])

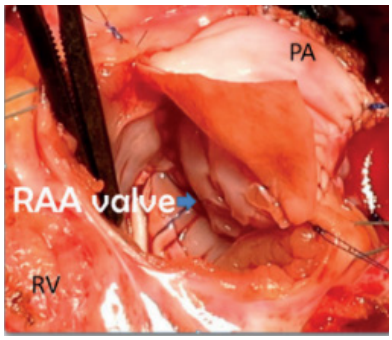

Figure 3 – Bicuspid neovalve (the figure was taken from [1]

was 14 mmHg. There was no pulmonary valve regurgitation, and only light tricuspid regurgitation was observed. The VSD patch was found to be closed tightly. The patient's ejection fraction was 50%. The patient was admitted to the intensive care unit (ICU) for two days and was extubated on the first day after surgery without complications. The patient was discharged after ten days. On the control echocardiogram before discharge, the gradient in the right ventricle to pulmonary artery (RV/PA) was 22 mmHg. There was a mild pulmonary regurgitation. The right part of the heart was dynamic with a reduction. The left

ventricular pumping and contractile function was satisfactory, with an ejection fraction (EF) of 60%.

Discussion

In the long term, the preservation of neopulmonary bicuspid valve function in Tetralogy of Fallot has a key value in paediatric cardiac surgery. Previously, numerous attempts have been made to preserve the pulmonary valve using both autologous and foreign tissue, but the results have been disappointing. Given that the use of the biological or mechanical valve in small children is not possible, this technique represents a potential solution to this problem. It is now possible to preserve the pulmonary valve for a long period of time, thanks to our colleagues from Iran, led by Amirghofran A. However, it should be noted that this method is not a panacea for all patients and that careful selection is required. In the case of an abnormally short, fused or absent appendage of the right atrium, this method is not applicable. In preparing the right atrial appendage (RAA), the following criteria must be met: the width of the RAA should be equal to half the circumference of the design annulus, and the height should be equal to the diameter of the design annulus. Additionally, the inner part of the right atrial appendage should be thoroughly cleaned of rough trabeculae. Given that the tissue of the right atrial appendage is autologous, anticoagulant therapy is not required. This surgical technique has no age restrictions and can be used in both 15-day-old babies and 57-year-old patients with tetralogy of Fallot. It is not limited to this condition and can also be applied in the treatment of pulmonary atresia type 1, Nikaidoh operation, common arterial trunk, and absent pulmonary valve syndrome [9-13].

Conclusion

It can be concluded that the use of the right atrial appendage as a bicuspid valve in the pulmonary position represents an alternative method for the correction of Tetralogy of Fallot with deformed, hypoplastic and aplasic pulmonary valve. The new technique is safe, easy to perform and the first successful results in Kazakhstan are promising. Given that the right atrial appendage is autologous tissue, there is a possibility of valve growth, which could represent a new branch in the improvement of surgical tactics in Tetralogy of Fallot. However, further follow-up is required to obtain data that are more reliable.

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