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REPEATED INTERVENTIONS AFTER TOTAL REPAIR IN PATIENTS WITH TETRALOGY OF FALLOT

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Tetralogy of Fallot occurs in 3.5 % of all congenital heart defects. The total correction consists in eliminating the obstruction of the right ventricular outflow tract using two main techniques: transannular patch and valve-sparing repair – non-transannular patch. The long-term results of 1352 radical corrections of tetralogy of Fallot performed at the Ukrainian Cardiac Center from 1999 to 2022. The the median age of patients at the time of surgery was 9.8 months. The use of transannular patch was 64.2 %, and Non-transannular patch – 35.8 %. The total mortality was – 3.46 %. Reinterventions were performed in 213 (16.5 %) patients. The most procedures were associated with pulmonary artery valve insufficiency (55.5 %) and right ventricular outflow tract obstruction (40.7 %). Analysis of risk factors showed that the use of transannular patch is an independent risk factor for repeated operations (r=0.726, p<0.001).

Key words: congenital heart defects, palliation, surgical correction, endovascular interventions, transannular plasty, valve-sparing surgery.

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ПОВТОРНІ ВТРУЧАННЯ ПІСЛЯ РАДИКАЛЬНОЇ КОРЕКЦІЇ ТЕТРАДИ ФАЛЛО

Тетрада Фалло зустрічається у 3,5 % всіх вроджених вад серця. Корекція даної вади: усунення обструкції вихідного тракту правого шлуночка, проводиться з використанням двох основних технік: трансанулярної латки і клапанозберігаючої техніки. У дослідженні проведено ретроспективний аналіз віддалених результатів 1352 радикальних корекцій тетради Фалло проведених у ДУ «Науково-практичний медичний центр дитячої кардіології та кардіохірургії МОЗ України» в період з 1999 по 2022 роки. Середній вік пацієнтів при хірургічному втручанні становив 12,7 місяців, медіана віку – 9,8 місяці. Застосування трансанулярної латки становило 64,2 %, а клапанозберігаючої техніки – 35,8 %. Рання післяопераційна летальність склала 2,36 %, а загальна летальність – 3,46 %. Повторні втручання проводились у 213 (16,5 %) пацієнтів. Більшість повторних операцій було пов'язано з недостатністю клапана легеневої артерії (55,5 %) та обструкцією вихідного тракту правого шлуночка (40,7 %). Аналіз факторів ризику, за результатами кореляційного аналізу, показав, що застосування клапанозберігаючої техніки є незалежним фактором ризику повторних операцій (r=0,726, p<0,001).

Ключові слова: вроджені вади серця, паліативні втручання, хірургічна корекція, ендоваскулярні втручання, трансанулярна пластика, клапанозберігаюча операція.

The work is a fragment of the initiative research project.

Tetralogy of Fallot (ToF) is a congenital heart defect that occurs in approximately 3.5 % of infants with cardiovascular diseases. Tetralogy of Fallot arises due to an anomaly in the development of the conus, with anterior deviation of the conotruncal septum, leading to the narrowing of the right ventricular outflow tract (RVOT), the formation of a large ventricular septal defect, and right ventricular (RV) hypertrophy.

A significant issue in the long-term follow-up period after surgical correction is the need for repeated interventions due to pulmonary valve stenosis or insufficiency [1, 2]. The frequency of reoperations in ToF may vary depending on numerous factors, including anatomical variations of the defect and the technique used in the primary surgery: valve-sparing technique, non-transannular patch (NTAP), or transannular patch (TAP) with incision of the pulmonary valve annulus [3]. Overall, statistics indicate that approximately 20–30 % of patients may require repeat surgical intervention within 10–15 years after the primary operation [4, 5].

The purpose of the study was to determine the risk factors for repeat interventions after surgical correction of Tetralogy of Fallot.

Materials and methods. This retrospective study presents the results of analysis of the long-term outcomes fot the 1,352 radical corrections in patients with ToF and double outlet right ventricle (tetralogy type), operated on between 1999 and 2022 at the Ukrainian Cardiac Center. The average age at the time of surgery was 12.7±44.3 (0.1–575 months), with a median age of 9.8 months. The average weight was 10.2±10.1 kg (1.9–90 kg), with a median weight of 10.1 kg. The number of male and female patients was 736 and 616, respectively. The different diagnoses and forms of Tetralogy of Fallot are present in Table 1.

In 236 patients (17.5 %), ToF was associated with a right aortic arch; in 185 patients (14 %), it was accompanied by a patent ductus arteriosus (PDA). Significant stenosis of one or both pulmonary artery

branches were detected in 163 patients (12 %). An additional left superior vena cava draining into the coronary sinus was found in 70 patients (5.1 %), while a complete vascular ring was identified in 71 patients (5.2 %). Among them, two patients had a right aortic arch with an aberrant left subclavian artery, and one had a circumflex aortic arch with an aberrant left subclavian artery. DiGeorge syndrome was diagnosed in 28 patients (2.1 %) out of 1,352, while Down syndrome was found in 54 patients (3.99 %).

Table 1

Anatomical variants in patients with Tetralogy of Fallot

Diagnosis	Number (n=)	Percentage (%)
ToF	970	71.8
DORV	322	23.8
TOF with absent pulmonary valve syndrome (APVS)	26	1.9
TOF with atrioventricular canal (AVC)	34	2.5
Total	1.352	100

ToF – Tetralogy of Fallot; DORV – Double Outlet Right Ventricle; ToF with APVS – Tetralogy of Fallot with absent pulmonary valve syndrome; ToF with AVC – Tetralogy of Fallot with atrioventricular canal.

Previous palliative interventions, including Blalock-Taussig (BTSh) or central systemic-to-pulmonary shunts, were performed in 173 patients (12.79 %) and previous balloon pulmonary valvuloplasty was conducted in 165 patients (12.2 %). RVOT obstruction relief using the TAP technique was performed in 64.13 % of patients (867 cases), while NTAP was used in 35.87% (485 cases).

Between 1999 and 2009, 468 radical corrections of ToF were performed, with a transannular patch used in 279 patients (59.62 %) and a valve-sparing technique in 189 patients (40.38 %). A similar ratio of these two methods was observed between 2010 and 2020. With an overall increase in the number of operations (884), the transannular technique was applied in 588 patients (66.5 %), while the valve-sparing technique was used in 296 patients (33.5 %) (Fig. 1).

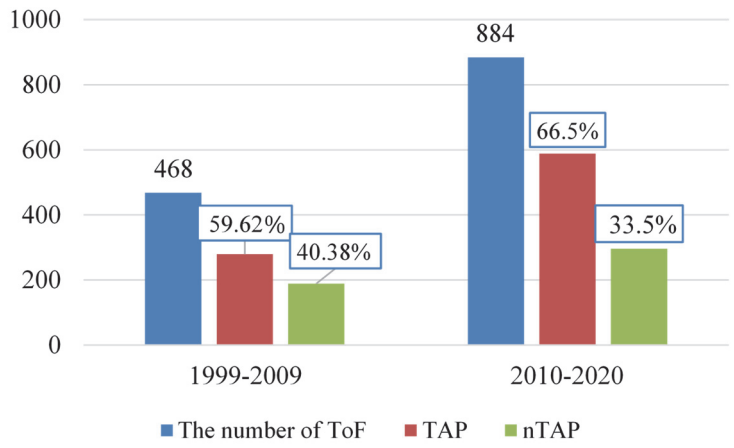

Fig. 1. The ratio of different techniques used for right ventricular outflow tract obstruction correction over the years.

For data analysis obtained during the study, IBM SPSS Statistics 21.0 and Microsoft Excel 2016 software were used. All calculations were performed with a confidence level of 95 %, meaning that differences between groups were considered statistically significant at $p < 0.05$. The study was conducted in accordance with the principles of the Helsinki Declaration. The study protocol was approved by the Local Ethics Committee of the institute. Informed consent was obtained from the parents of the children for the research.

Results of the study and their discussion. Early postoperative mortality was recorded in 32 patients (2.36 %). The long-term follow-up period was 96.3±62.7 months (3–241 months). Late mortality was observed in 14 patients (1.1 %). The overall mortality rate was 3.46 %. The long-term follow-up was assessed in 1.284 (94.92 %) patients, as 22 (1.62 %) patients were lost to follow-up.

Repeated interventions, both surgical and endovascular, were performed in 213 (16.5 %) patients, with a total of 305 (23.8 %) procedures conducted. Endovascular reinterventions were performed in 57 patients, with a total of 116 (8.6 %) procedures, including: 59 pulmonary valve balloon dilations (PVB) in 34 patients; 36 PVB with stenting of pulmonary artery branches in 14 patients; 21 balloon dilations of pulmonary artery branches in 9 patients.

Repeat surgical interventions were performed in 156 patients, with a total of 189 surgeries, accounting for 14.8 % of all primary corrections. The average time between radical correction of ToF and reoperation was 79.7±59.7 months (8.17–311 months). The reasons for late repeat surgical interventions were: pulmonary valve insufficiency in 105 cases (55.5 %); right ventricular outflow tract obstruction in 77 cases (40.7 %); infective endocarditis in 2 cases (1 %); pulmonary artery thromboembolism in 1 case (0.5 %).

The types of reoperations are presented in Table 2. The majority of repeat surgeries for ToF were due to pulmonary valve insufficiency – 105 cases (55.5 %), requiring the implantation of a right ventricle to pulmonary artery conduit. Correction of right ventricular outflow tract obstruction in 77 cases (40.7 %) included: transannular patch and infundibulectomy in 45 patients; transannular patch with monocusp valve in 29 patients; replacement of a calcified transannular patch in 3 patients. Other reoperations included closure of significant residual ventricle septal defect shunts, removal of vegetations, and plastic reconstruction of pulmonary artery branches.

Table 2

Types of reoperations in Tetralogy of Fallot

Type of reoperations	n=189	%
Conduit implantation	105	55.5
Correction of RVOT obstruction	77	40.7
VSD residual shunt closure	4	2.3
Endocarditis treatment	2	1
Pulmonary artery branch reconstruction	1	0.5

Hospital mortality for repeat surgeries was 0.5 % (1 patient). The patient underwent reoperation six years after surgical correction due to tubular stenosis of the central and peripheral pulmonary artery branches. The cause of death was acute right ventricular failure with suprasystemic pressure.

During the surgical stage, cardiac injury and acute bleeding occurred in 24 cases (12.6 %) during resternotomy, none of which were fatal.

Early postoperative complications occurred in 16 patients (10.25 %), including: acute bleeding requiring reoperation in 11 patients (7.05 %); acute renal failure requiring replacement therapy in 3 patients (1.9 %); acute cerebrovascular accident in 2 patients (1.3 %).

A regression correlation analysis used to assess the relationship between surgical correction methods of ToF and reoperations showed that a significant independent risk factor for repeat surgical interventions was transannular patch repair ($r=0.726$ (0.32–0.85), $p < 0.001$).

Surgical correction in Tetralogy of Fallot (ToF) involves ventricular septal defect (VSD) closure and right ventricular outflow tract (RVOT) reconstruction [6]. Residual RVOT obstruction after radical defect correction leads to right ventricular hypertrophy, which is a risk factor for reoperations and increases the likelihood of hospital morbidity and late mortality [7]. Meanwhile, significant pulmonary valve insufficiency can be well tolerated by patients even after radical correction. Mild pulmonary valve insufficiency, with a regurgitation fraction of approximately 40 %, does not cause serious problems over an extended period, as the pulmonary microcirculatory system is capable of compensating for this defect. Therefore, minor pulmonary valve regurgitation is considered less dangerous than prolonged right ventricular dilation, which leads to dysfunction [8].

The classical approach to radical ToF correction involves extended ventriculotomy with transventricular VSD closure and transannular patching. This technique includes the excision of RVOT obstruction via right ventricular ventriculotomy, followed by VSD closure. This method is widely used worldwide due to its effectiveness, but it has drawbacks: the necessity of right ventricular ventriculotomy may lead to ischemia and akinesia of the right ventricular anterior wall, as well as progressive pulmonary valve insufficiency due to infundibular dysfunction.

In the study group of 1,352 patients, RVOT obstruction was corrected using two primary techniques: transannular patching (TAP) and non-transannular patching (NTAP). TAP was used in 64.2 % of cases, while NTAP was applied in 35.8 %. Between 1999 and 2009, the use of TAP was 59 %, whereas from 2010 to 2020, it increased to 66.5 %. Early postoperative mortality was 2.36 %, and overall mortality at the time of long-term follow-up was 3.46 %, indicating a high success rate of surgeries. The average follow-up period was 96.3 months.

Repeat surgical and endovascular interventions were performed in 213 patients (16.5 %). The majority of repeat surgeries were associated with pulmonary valve insufficiency (55.5 %) and RVOT obstruction (40.7 %). Endovascular procedures included pulmonary valve balloon dilations (PVB) and stenting. Retrospectively, the primary factor increasing the likelihood of repeat surgeries was the use of transannular patching (TAP). Correlation analysis showed that TAP is an independent risk factor for repeat surgeries ($r=0.726$, $p<0.001$). It was also found that surgeries performed before 2010 had a higher risk of requiring reoperations.

Radical ToF correction is a complex and highly specialized surgical intervention that requires careful selection of the method based on patient age, disease stage, and individual cardiac anatomy.

Specifically, in newborns and young children with disease symptoms, new options have emerged to delay surgical intervention: ductal stenting, pulmonary valve balloon dilation, and RVOT stenting. Palliative interventions in the neonatal period allow for delaying surgery and reducing the risk of repeat interventions after surgical correction [9, 10]. Despite the successful outcomes of traditional methods, new pulmonary valve-sparing techniques may reduce the risks of long-term insufficiency and improve long-term outcomes. However, these methods still require further research to confirm their long-term effectiveness and safety [10, 11].

Conclusions

1. Tetralogy of Fallot is a congenital heart defect with a high risk of repeat interventions, both endovascular and surgical – 305 (23.5 %) cases out of 1.284 patients who survived surgical correction and were followed for a 20-year period.

2. The number of reoperations increases with longer follow-up periods; after 15 years, their rate reaches 25–30 % of the number of primary surgeries.

3. Reoperations after ToF correction are safe, with a reoperation mortality rate of 0.5 % and a low rate of early complications (8.4 %).

4. The transannular path repair (r=0.726 (0.32-0.85), p<0.001) was identified as a significant independent risk factor for repeat surgical interventions, pulmonary valve-preserving correction of ToF should be considered the preferred method.

5. The valve-sparing technique for RVOT obstruction relief in ToF correction leads to a lower risk of pulmonary valve insufficiency and reduces the need for conduit implantation in the long-term follow-up period.

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