

The role of early unifocalization in the correction of pulmonary atresia with ventricular septal defect: the retrospective analysis

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Received 28 November 2024. Revised 25 September 2025.
Accepted 30 January 2026.

How to cite: Amansakhatova E.N., Kulyabin Yu.Yu., Voitov A.V., Nichay N.R., Arkhipov A.N., Soynov I.A. The role of early unifocalization in the correction of pulmonary atresia with ventricular septal defect: the retrospective analysis. *Patologiya krovoobrashcheniya i kardiokirurgiya = Circulation Pathology and Cardiac Surgery*. 2026;30(2):45-54. (In Russ.)
<https://doi.org/10.21688/1681-3472-2026-2-45-54>

Funding

The research was carried out within the framework of the state assignment of the Ministry of Health of the Russian Federation (No. 124022500251-0).

Conflict of interest

The authors declare no conflict of interest.

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Abstract

Introduction: The presence of multifocal pulmonary blood flow in pulmonary atresia with ventricular septal defect plays a critical role in determining the strategy for surgical correction. Early unifocalization of the pulmonary arterial bed facilitates achievement of complete repair with physiological right ventricular pressure.

Objective: To evaluate the effectiveness of pulmonary artery reconstruction with unifocalization to achieve radical correction in pulmonary artery atresia with interventricular defect and multifocal pulmonary flow.

Methods: This study involved 25 patients with pulmonary atresia and ventricular septal defect types B and C (according to the classification of C. Tchervenkov), who underwent unifocalization of the pulmonary bed. In accordance with age at the time of correction, all patients were divided into two groups: Group EU (early unifocalization, $n = 11$) – patients operated on at the age of up to 1 year, Group LU (late unifocalization, $n = 14$) – patients operated on at the age of over 1 year. The primary endpoint was successful complete repair with maintenance of right ventricular pressure at less than two-thirds of systemic pressure. The secondary endpoints included the rates of reintervention, freedom from reoperation, incidence of residual pulmonary hypertension, and rate of adverse outcomes.

Results: The age of patients in the groups with early and late unifocalization of the pulmonary bed was 6.0 (0; 11) and 90.5 (13; 132) months. Pulmonary hypertension in the preoperative period was diagnosed in 1 patient (9 %) in the EU group, and in 11 patients (79 %) in the LU group ($p = 0.001$). Radical correction and right ventricular pressure less than 2/3 of the systemic pressure were achieved more often in patients in group EU than in group LU (73 % vs. 29 %, $p = 0.04$ and 89 % vs. 38 %, $p = 0.03$). Repeated transcatheter interventions and surgical redo procedures were required more often in patients from group LU (85 %, $p = 0.02$, 85 %, $p = 0.007$). There were no statistically significant differences in early and long-term mortality rates between the groups ($p = 1.0$).

Conclusion: Early unifocalization of pulmonary blood flow allows achieving optimal parameters for achieving radical correction of the defect. After radical correction, most patients who underwent unifocalization at an early age retain low pressure in the right ventricle in the long term and are less susceptible to repeated interventions.

Keywords: pulmonary atresia with ventricular septal defect; congenital heart disease; unifocalization of the pulmonary arteries

Ранняя унифокализация при коррекции атрезии легочной артерии с дефектом межжелудочковой перегородки: ретроспективный анализ

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Поступила в редакцию 28 ноября 2024 г.
Исправлена 25 сентября 2025 г. Принята к печати 30 января 2026 г.

Цитировать: Амансахатова Е.Н., Кулябин Ю.Ю., Войтов А.В., Ничай Н.Р., Архипов А.Н., Сойнов И.А. Ранняя унифокализация при коррекции атрезии легочной артерии с дефектом межжелудочковой перегородки: ретроспективный анализ. *Патология кровообращения и кардиохирургия*. 2026;30(2):45–54.
<https://doi.org/10.21688/1681-3472-2026-2-45-54>

Финансирование

Работа выполнена в рамках государственного задания Министерства здравоохранения Российской Федерации (№ 124022500251-0).

Конфликт интересов

Авторы заявляют об отсутствии конфликта интересов.

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Аннотация

Актуальность. Наличие мультифокального легочного кровотока при атрезии легочной артерии и дефекте межжелудочковой перегородки играет решающую роль в выборе тактики хирургической коррекции. Ранняя унифокализация легочного русла способствует достижению радикальной коррекции порока с физиологическим давлением в правом желудочке.

Цель. Оценить эффективность реконструкции легочного русла с унифокализацией для достижения радикальной коррекции при атрезии легочной артерии с дефектом межжелудочковой перегородки и мультифокальным легочным кровотоком.

Методы. В данном исследовании приняли участие 25 пациентов с атрезией легочной артерии и дефектом межжелудочковой перегородки типов В и С (по классификации С. Tchervenkov), которым выполнялась унифокализация легочного русла. В соответствии с возрастом во время коррекции все пациенты были разделены на две группы: группа с ранней унифокализацией (РУ, $n = 11$) – пациенты, оперированные в возрасте до 1 года, группа с поздней унифокализацией (ПУ, $n = 14$) – пациенты, оперированные в возрасте старше 1 года. Первичной конечной точкой исследования являлось достижение радикальной коррекции с поддержанием давления в правом желудочке на уровне менее 2/3 от системного давления. Вторичные точки включали частоту повторных эндоваскулярных вмешательств, свободу от реопераций, частоту резидуальной легочной гипертензии, частоту неблагоприятного исхода.

Результаты. Возраст пациентов в группах с ранней и поздней унифокализацией легочного русла составил 6,0 (0; 11) и 90,5 (13; 132) месяца. Легочная гипертензия в предоперационном периоде была диагностирована в группе РУ у 1 пациента (9 %), а в группе ПУ – у 11 пациентов (79 %) ($p = 0,001$). Радикальная коррекция и давление в правом желудочке менее 2/3 от системного чаще достигались у пациентов в группе РУ, чем в группе ПУ (73 % vs 29 %, $p = 0,04$ и 89 % vs 38 %, $p = 0,03$). Повторные транскатетерные и открытые хирургические вмешательства требовались чаще пациентам из группы ПУ (85 %, $p = 0,02$, 85 %, $p = 0,007$). В показателях ранней и отдаленной летальности между группами не было статистически значимых различий ($p = 1,0$).

Заключение. Результаты исследования дают возможность предположить, что ранняя унифокализация легочного кровотока позволяет обеспечить оптимальные показатели для достижения радикальной коррекции порока. После радикальной коррекции у большинства пациентов, которым была проведена унифокализация в раннем возрасте, сохраняется низкое давление в правом желудочке в отдаленном периоде, и они в меньшей степени подвержены повторным вмешательствам.

Ключевые слова: атрезия легочной артерии с дефектом межжелудочковой перегородки; врожденные пороки сердца; унифокализация легочных артерий



Introduction

Pulmonary atresia (PA) with ventricular septal defect (VSD) is often accompanied by the development of major aortopulmonary collateral arteries (MAPCAs). The presence of MAPCAs in PA with VSD is associated with higher mortality, and ten-year survival rates in the natural history of this condition do not exceed 10 % [1–3]. The presence of MAPCAs creates a specific physiological course characterized by the rapid development of pulmonary hypertension, which complicates the choice of surgical strategy for PA with VSD [4; 5]. The main condition for achieving complete repair of the congenital heart defect is low pulmonary vascular resistance with normal arborization of the pulmonary arteries [4; 6]. In non-communicating MAPCAs, the primary approach to achieving physiological pulmonary blood flow is unifocalization of the pulmonary circulation [7; 8].

The aim of our study was to evaluate outcomes of early unifocalization of the pulmonary blood flow to achieve adequate complete repair of PA with VSD.

Methods

In this retrospective analysis, 44 patients with PA/VSD and multifocal pulmonary blood flow who underwent surgery at the Department of Congenital Heart Defects of the E.N. Meshalkin National Medical Research Center between January 2018 and July 2024 were included. The analysis included children with PA/VSD types B and C according to the Tchervenkov classification who underwent surgical correction of the defect with unifocalization of the pulmonary bed. Of the 44 patients, 25 children were selected for further analysis and divided into two groups according to the age at which neopulmonary arteries reconstruction was performed: Group I – early unifocalization (EU), under 1 year of

age (11 patients); Group II – late unifocalization (LU), performed at an age older than 1 year (14 patients) (Fig. 1).

The inclusion and exclusion criteria for the study

- Inclusion criteria:**
- Patient age at the time of unifocalization: under 1 year in the EU group and over 1 year in the LU group.
 - PA with VSD types B and C according to the Tchervenkov classification;
 - Primary surgical correction.

Exclusion criteria:

- Multiple congenital malformations, low body weight;
- Patients who underwent repeat correction at another center;
- Bronchopulmonary developmental anomalies;
- Refusal to participate in the study.

Endpoints

Primary Endpoint: achievement of complete repair with preservation of right ventricular pressure below 2/3 of systemic pressure.

Secondary Endpoints: frequency of repeat endovascular interventions, freedom from reoperation, frequency of residual pulmonary hypertension, frequency of adverse outcomes.

All patients underwent contrast-enhanced multislice computed tomography of the heart and great vessels as well as transvenous cardiac catheterization with angiopulmonography and aortography to assess the anatomy and physiology of the pulmonary bed and the presence of multifocal pulmonary blood flow. In the mid-term period (mid-term outcomes are defined as events occurring after hospital discharge), all patients underwent repeat control cardiac catheterization with angiopulmonography and tensiometry to assess

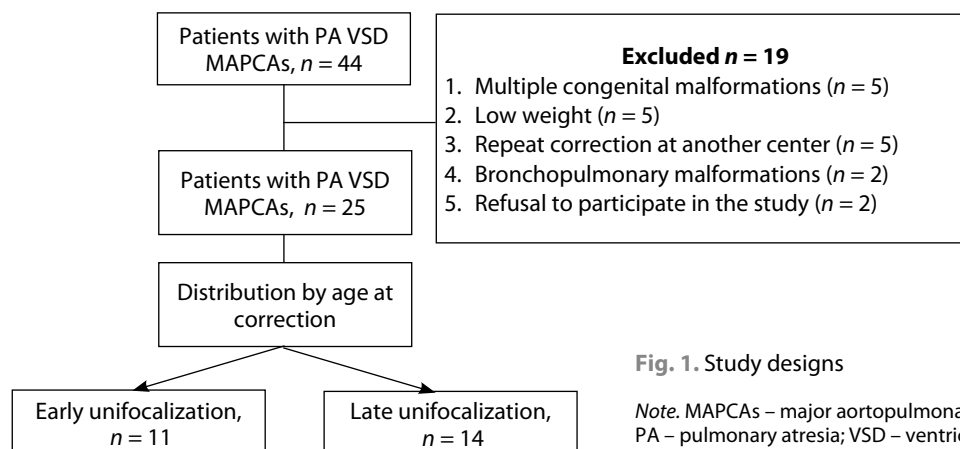


Fig. 1. Study designs

Note. MAPCAs – major aortopulmonary collateral arteries; PA – pulmonary atresia; VSD – ventricular septal defect.

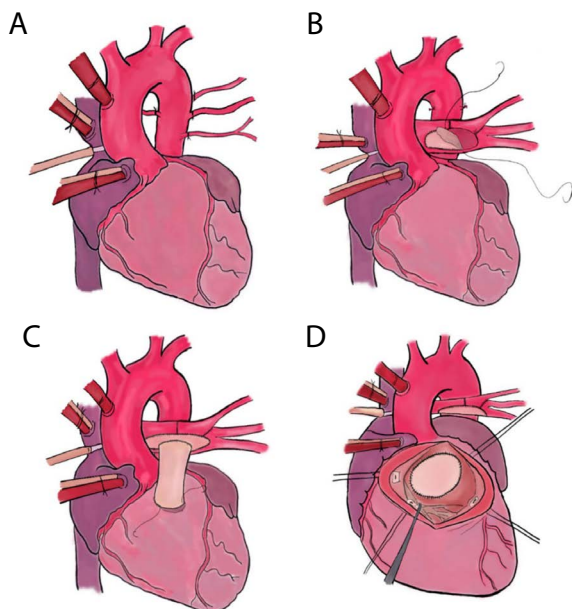


Fig. 2. Complete repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: A – ligation or clipping of MAPCAs; B – unifocalization of the pulmonary vascular bed; C – reconstruction of the RVOT; D – closure of the VSD

Note. MAPCAs – major aortopulmonary collateral arteries; RVOT – right ventricular outflow tract; VSD – ventricular septal defect.

pulmonary circulation physiology and neopulmonary artery anatomy at 1 year after surgery. When measuring pressure in the right ventricle (RV) and the neopulmonary bed, comparable values were obtained in the absence of a ventricular septal communication (including a fistula in the patch).

The indications for surgical correction with unifocalization were the presence of multifocal pulmonary blood flow with non-communicating MAPCAs, significant MAPCA stenoses, desaturation below 80 %, or large MAPCAs with the development of signs of heart failure. Unifocalization of the pulmonary arteries was performed from the age of 4 months in the presence of non-communicating MAPCAs with a diameter greater than 2 mm. The decision to close the VSD was made based on intraoperative assessment of the neopulmonary bed anatomy and direct tensiometry in the RV after unifocalization. When unifocalized MAPCAs were small and RV pressure exceeded 2/3 of systemic pressure, palliative correction was performed with creation of a shunt between the RV and the neopulmonary collector, leaving a non-restrictive VSD.

Surgical technique

Unifocalization was performed via a median sternotomy approach using cardiopulmonary bypass (CPB) in all cases (Fig. 2). Prior to patient heparinization, mobilization of the pulmonary branches (if present) and all MAPCAs was performed down to their division into segmental branches. After initiation of CPB, all MAPCAs were clipped at the aortic origin (Fig. 2, A). The collaterals were sequentially connected to each other and to the native pulmonary bed (if present) using multiple anastomoses, followed by extended patch plasty with cryopreserved pulmonary homograft tissue or autopericardium (Fig. 2, B). After creation of a single pulmonary collector, neopulmonary arteries were assessed, followed by the creation of an anastomosis between the conduit and the collector. For complete repair, a valved xenograft conduit Contegra (Medtronic, USA) or a cryopreserved venous homograft of a calculated diameter was implanted. In the case of palliative procedure, a non-valved synthetic graft was implanted (Fig. 2, D). Next, aortic occlusion was performed, and Bretschneider's cardioplegic solution (Custadiol, Germany) was used for myocardial protection. The right ventricular outflow tract was opened in an avascular area, partial resection of the conal septum was performed, after which the VSD was closed with a xenopericardial patch (Fig. 2, C). Through a right atrial approach, a decompression fistula was additionally created in the interatrial septum with a diameter of 4 mm. After discontinuation of CPB, direct tensiometry was performed in the RV and pulmonary artery (PA). If high pressure (more than 2/3 of systemic pressure) persisted, CPB was resumed for volume unloading and a fistula was created in the ventricular patch under induced ventricular fibrillation.

Statistical Analysis

Statistical analysis was performed using Stata 13 software (StataCorp LP, College Station, USA). The normality of distribution of the variables was assessed using the Shapiro–Wilk test. Continuous variables are presented as median (25th; 75th percentile; minimum and maximum values in the sample). Categorical variables are presented as numbers *n* (%). For intergroup comparison of continuous data, the Mann–Whitney test was used. For comparison of groups with categorical data, the χ^2 test or Fisher's exact test was used. For the analysis of qualitative variables, Pearson's χ^2 test was applied; in cases where less than 25 % of expected frequencies exceeded 5, Fisher's exact test was used. Comparative analysis of five-year freedom from repeat open interventions was performed using the log-rank test. A significance level of $p < 0.05$ was adopted.

Results

Table 1. Baseline and demographic characteristics of the patients

Characteristics	Early unifocalization, <i>n</i> = 11	Late unifocalization, <i>n</i> = 14	<i>p</i>
Age, months	6 (4; 8)	90.5 (25; 96)	0.001
Weight, kg	5.2 (4; 8)	15.2 (7; 18)	0.001
Pulmonary atresia with ventricular septal defect, type C, <i>n</i> (%)	7 (63)	7 (50)	0.680
Pulmonary hypertension, <i>n</i> (%)	1 (9)	11 (79)	0.001
SpO ₂ , %	82 (80; 90)	80 (70; 80)	0.460
Concomitant non-cardiac pathology, <i>n</i> (%)	8 (73)	8 (57)	0.670
Palliative procedures before unifocalization, <i>n</i> (%)	4 (36)	9 (64)	0.230

Note. Data are presented as median (25th; 75th percentile) or as numbers *n* (%).

The baseline and demographic characteristics of patients are summarized in Table 1.

According to selective tensiometry, in the preoperative period, pulmonary hypertension was diagnosed in only 1 patient (9 %) in the EU group, compared to 11 patients in the LU group (79 %) ($p = 0.001$). In the LU group, 11 patients (79 %) underwent palliative procedures on the pulmonary arteries and MAPCAs before unifocalization.

At the time of hospitalization, many patients had concomitant diagnoses involving pathology of other organs, including systemic conditions, such as central nervous system pathology (previous stroke), DiGeorge syndrome, cytomegalovirus infection, and polycystic kidney disease.

Complete repair was achieved in 73 % of patients in the EU group and in 29 % of patients in the LU group ($p = 0.04$) (Tabl. 2). The majority of patients in each group underwent extended pulmonary branch plasty due to the presence of peripheral stenoses and small-diameter MAPCAs. (73 % vs 79 %, $p = 0.07$).

In-hospital mortality did not differ significantly between the groups and was 9 % ($n = 1$) in the EU group and 7 % in the LU group ($n = 1$) ($p > 0.99$). Extracorporeal therapy (extracorporeal membrane oxygenation, ECMO) was required in 9 % ($n = 1$) of patients in the EU group and in 14 % ($n = 2$) of patients in the LU group ($p > 0.99$).

One patient in the EU group developed acute occlusion of the left pulmonary artery on postoperative day 2, leading to right ventricular failure. ECMO was initiated,

Table 2. Intraoperative and early postoperative outcomes

Characteristics	Early unifocalization, <i>n</i> = 11	Late unifocalization, <i>n</i> = 14	<i>p</i>
Complete repair, <i>n</i> (%)	8 (73)	4 (29)	0.040
CPB time, min	215 (137; 258)	225 (174; 254)	0.600
Aortic cross-clamp time, min	42 (39; 52)	39 (37; 62)	0.850
Extended pulmonary artery branch plasty, <i>n</i> (%)	8 (73)	11 (79)	>0.999
Delayed chest closure, <i>n</i> (%)	5 (45)	3 (21)	0.380
SpO ₂ , %	97 (90; 96)	92 (85; 97)	0.200
Hospital mortality, <i>n</i> (%)	1 (9)	1 (7)	>0.999
ECMO, <i>n</i> (%)	1 (9)	2 (14)	>0.999
Complications			
Bronchial stenoses, <i>n</i> (%)	3 (27)	3 (21)	>0.999
Branch occlusion, <i>n</i> (%)	1 (9)	3 (21)	0.600
Infections, <i>n</i> (%)	2 (18)	1 (7)	0.560

Note. CPB – Cardiopulmonary bypass; ECMO – extracorporeal membrane oxygenation. Data are presented as median (25th; 75th percentile) or as numbers *n* (%).

Table 3. Characteristics of the mid-term postoperative period (for patients discharged from the hospital)

Characteristics	Early unifocalization, <i>n</i> = 10	Late unifocalization, <i>n</i> = 13	<i>p</i>
Long-term mortality, <i>n</i> (%)	1 (9)	0 (0)	0.170
Follow-up, months	16 (8; 36)	21 (8; 36)	–
Endovascular reinterventions within 1 year after surgery, <i>n</i> (%)	3 (33)	11 (85)	0.020
Three-year freedom from reoperation, <i>n</i> (%)	0 (0)	10 (82)	0.020
Pressure RV < 2/3, <i>n</i> (%)	8 (89)	5 (38)	0.030
FAC RV, %	45 (42; 50)	42 (38; 42)	–
Peak pressure gradient across PV, mmHg	12 (5; 18)	6 (5; 18)	–
TR:			
grade 0–1, <i>n</i> (%)	7 (78)	9 (69)	–
grade 1–2, <i>n</i> (%)	2 (22)	4 (31)	–

Note. RV – right ventricle; TR – tricuspid regurgitation; FAC – Fractional Area Change; PV – pulmonary valve. Data are presented as median (25th; 75th percentile) or as numbers *n* (%).

and transcatheter recanalization with stenting of the occluded segment was performed. The patient died from progressive multiorgan failure on postoperative day 18. In the LU group, one patient developed reperfusion lung injury on the first postoperative day, resulting in severe respiratory failure. Veno-arterial ECMO was initiated; however, the development of infectious complications

and pneumonia was the primary cause of death on postoperative day 22.

In the early postoperative period, bronchial obstruction with bronchial stenosis was diagnosed in three patients (27 %) in the EU group and in three patients (21 %) in the LU group, requiring prolonged mechanical ventilation.

Fig. 3. Five-year freedom from repeat open interventions (Kaplan – Meier method) in the two groups

Note. LU – late unifocalization;
EU – early unifocalization.

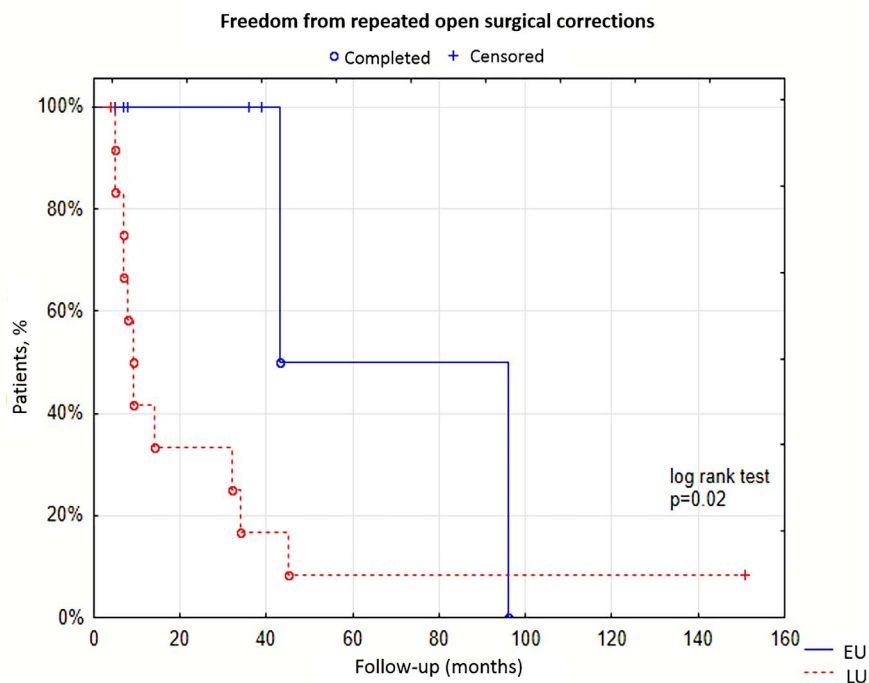
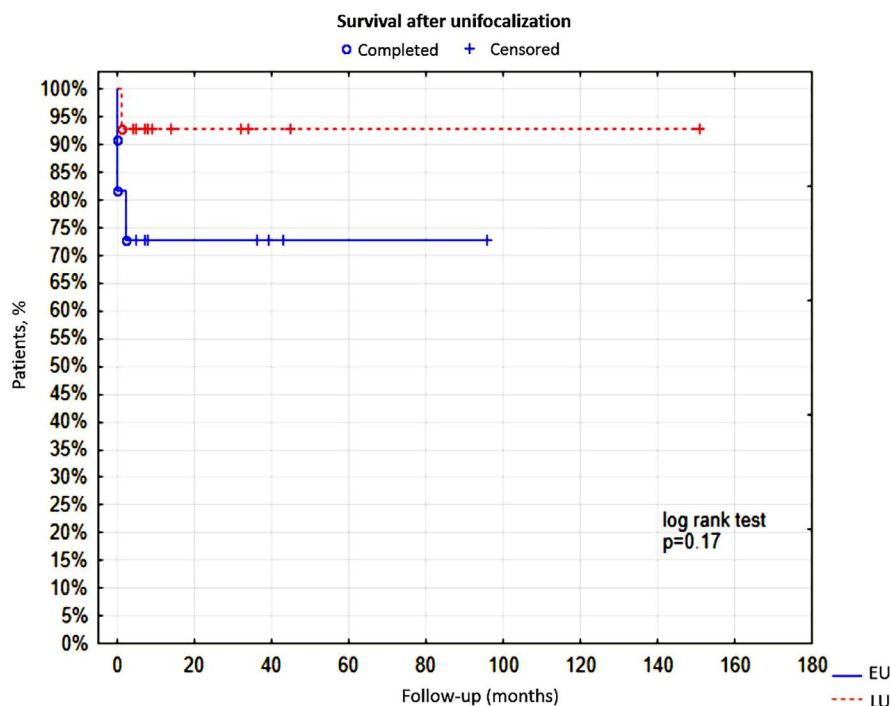


Fig. 4. Kaplan – Meier method: assessment of five-year survival in two groups

Note. LU – late unifocalization;
EU – early unifocalization.



The incidence of acute occlusion of unifocalized branches did not differ between the groups (9 % vs 21 %, $p = 0.600$).

In the majority of patients in the EU group, low right ventricular pressure was maintained according to control angiopulmonography in the mid-term period, in contrast to patients in the LU group (89 % vs 38 %, $p = 0.030$). Mid-term postoperative characteristics are shown in Table 3.

During the first year of follow-up, repeat endovascular interventions, including balloon angioplasty or stenting of pulmonary artery branches, were performed in 3 patients (33 %) in the EU group and in 11 patients (85 %) in the LU group (Log rank test, $p = 0.02$). The three-year freedom from repeat open surgical correction was 100 % in the EU group and 18 % in the LU group. (Log rank test, $p = 0.020$) (Fig. 3).

The maximum follow-up period was 5 years. One patient in the EU group died in the long-term period due to progressive multiorgan failure secondary to viral pneumonia (9 %); there was no long-term mortality in the LU group ($p = 0.440$). However, no statistically significant difference in survival was found between the groups: at 5 years after surgical correction, overall freedom from mortality was 72 % in the EU group and 92 % in the LU group (Log rank test, $p = 0.170$) (Fig. 4).

Discussion

PA/VSD with impaired arborization of the neopulmonary arteries leads to mosaic lung perfusion. Such patients are at high risk for developing pulmonary hypertension (systolic pulmonary artery pressure >30 mmHg) and have high mortality during the natural history of the defect (since aortopulmonary collaterals are prone to stenosing), reaching up to 90 % at ten years [3; 9–12]. Surgical repair strategies vary between centers due to the technical complexity of the procedure and the unpredictable course of the postoperative period [13–19]. The main aim of surgical repair of PA/VSD is to preserve RV function while achieving physiological pressure in the pulmonary circulation. Unifocalization of the pulmonary arteries enables physiological uniform pulmonary blood flow with the potential for pulmonary bed growth. Nevertheless, long-standing MAPCAs lead to the development of mosaic pulmonary hypertension and alterations in the pulmonary artery wall structure, with the risk of being unable to achieve complete repair [5; 20; 21]. According to our study, only 29 % of patients who underwent unifocalization at an older age achieved complete repair, while 85 % required repeated endovascular and open interventions to relieve stenoses of the neopulmonary arteries. These results indicate not only a rapid change in physiology but also the development of multiple deformities of the pulmonary branches, which complicates further patient rehabilitation. To prevent the

development of pulmonary hypertension and distortion of MAPCAs, centers with extensive experience in the surgical repair of this group of patients recommend performing correction with unifocalization at an age no older than 1 year [22; 23]. According to our data, in 89 % of children after early unifocalization, RV pressure levels allowing complete repair were achieved. On the other hand, the group of younger children may be at increased risk of complications in the early postoperative period, including surgical complications, airway obstruction requiring prolonged mechanical ventilation, as well as the development of infectious complications, which were the cause of death in one patient in the early unifocalization group. Among the most significant complications are acute thrombosis or deformation of the unifocalized branches due to their small diameter and tortuous course (retroesophageal, retrobronchial). The complex anatomy of MAPCAs results in a high probability of neopulmonary branch deformation after anastomosis creation, regardless of extended plasty. Such patients typically require emergency recanalization with stenting of the occluded segment to avoid impairment of pulmonary perfusion and worsening of right ventricular failure. In our patient cohort, there were no differences between the groups in the frequency of branch occlusions requiring urgent reinterventions or ECMO initiation.

Short-term survival rates in large cardiac surgery centers with extensive experience in unifocalization reach 90 % [5; 24], with low long-term mortality during follow-up periods of up to 10 years [23]. In our cohort, one younger patient and one patient older than one year died during the hospital stage. Given the small sample size and the development of specific complications with subsequent ECMO initiation in these patients, survival in our experience is comparable to that in centers performing neopulmonary arteries rehabilitation in patients with PA/VSD.

Most centers prefer to perform neopulmonary arteries reconstruction after the age of 1.5 years; however, in 2009, a group of authors led by F. Hanley first published results indicating more effective unifocalization in younger children [25]. The authors demonstrate that performing unifocalization at the age of 4–6 months not

only preserves right ventricular function and prevents the development of pulmonary hypertension but also reduces the risk of subsequent neopulmonary arteries distortion [26; 27]. The «Stanford strategy» is currently the most preferred approach, as it demonstrates excellent results in both the early and late postoperative periods; however, the technical complexity of early unifocalization limits its widespread adoption among cardiac surgery centers [28]. According to Watanabe et al., complete repair was achieved in all 27 patients who underwent unifocalization at an early age, with RV pressure remaining within physiological limits. During long-term follow-up, 2 patients died due to the development of an infectious-inflammatory process [27]. Our results also demonstrate that performing unifocalization in patients older than 1 year leads to a greater need for repeat surgery for peripheral stenoses of the neopulmonary arteries, as well as to persistent high pulmonary artery pressure and a high risk of right ventricular failure after complete repair. Moreover, the majority of patients already had pulmonary hypertension (79 %) at the time of hospitalization, which complicates the postoperative course and persists for a long time in the long term, despite therapy and the absence of anatomical obstruction. Thus, in 10 out of 14 patients in our cohort, systemic pressure in the RV persisted without the possibility of performing complete repair of the defect.

Study limitations

Since the present study was conducted at a single center, the sample size may limit the significance of the obtained results.

Conclusion

The results of the study suggest that early unifocalization of pulmonary blood flow enables more effective achievement of optimal pulmonary circulation physiology parameters for complete repair while preserving adequate right ventricular workload. Furthermore, early unifocalization reduces the frequency of both repeat endovascular and surgical repair in the long-term period.

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