

Surgical management of congenital heart diseases in the Russian Federation: national registry data for 2024

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Received 9 February 2026. Revised 24 February 2026.
Accepted 2 March 2026.

How to cite: Soynov I.A., Kim A.I., Movsesyan R.R., Teplov P.V., Miller A.Yu., Kulyabin Yu.Yu., Avramenko A.A., Gorbaticov K.V., Gorbatykh A.V., Nalimov K.A., Abramyan M.A., Boriskov M.V., Kovalev S.A., Svobodov A.A., Arkhipov A.N., Krivoshchekov E.V., Amansakhatova E.N., Tarasyuk E.S. Surgical Management of Congenital Heart Diseases in the Russian Federation: National Registry Data for 2024. *Patologiya krovoobrashcheniya i kardiokhirurgiya = Circulation Pathology and Cardiac Surgery*. 2026;30(1):5-25. (In Russ.) <https://doi.org/10.21688/1681-3472-2026-1-5-25>

Funding

The study did not have sponsorship.

Conflict of interest

The authors declare no conflicts of interest.

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Abstract

Introduction. Congenital heart diseases (CHDs) remain a leading cause of infant mortality and disability, underlining their high medico-social significance. In the context of rapid technological advancements, evolving demographic indicators, and improvements in prenatal diagnostics, systemic monitoring of treatment outcomes at the national level has become critically important. The national registry facilitates an objective assessment of current cardiac surgical care effectiveness nationwide, helps identify regional trends, and determines priority areas for the implementation of modern hybrid and endovascular protocols.

Objective. To evaluate the current status and outcomes of surgical treatment for congenital heart diseases (CHD) in the Russian Federation based on the 2024 national multicenter registry data.

Methods. A retrospective analysis of 14,640 surgical procedures performed across 31 cardiac surgery centers was conducted. The RACHS II scoring system was utilized for risk stratification. The study assessed hospital mortality rates, pathology distribution, and the efficacy of various surgical strategies.

Results. The overall hospital mortality rate was 1.42%. Endovascular procedures accounted for a significant proportion (38.4%) of interventions. In the interventional arrhythmia segment ($n = 1,342$), zero mortality was achieved. The highest risk remains in the neonatal group (6.6% mortality). Analysis of palliative care revealed the superiority of the hybrid approach: ductal stenting in duct-dependent patients showed half the mortality rate (4.95%) compared to the surgical systemic-to-pulmonary artery shunt (9.74%). Complex reconstructions (Norwood procedure) remain a high-risk area.

Conclusion. The national pediatric cardiac surgery registry demonstrates high efficacy and safety for routine interventions. There is a distinct trend to-

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wards replacing open surgery with endovascular techniques. Centralization of care for critical CHDs in expert centers and the adoption of hybrid technologies are key factors for further improving outcomes.

Keywords: congenital heart disease; endovascular treatment; hospital mortality; national registry; pediatric cardiac surgery



Хирургическое лечение врожденных пороков сердца в Российской Федерации: данные национального регистра за 2024 год

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Поступила в редакцию 9 февраля 2026 г.
Исправлена 24 февраля 2026 г. Принята к печати 2 марта 2026 г.

Цитировать: Соинов И.А., Ким А.И., Мовсесян Р.Р., Теплов П.В., Миллер А.Ю., Кулябин Ю.Ю., Авраменко А.А., Горбатов К.В., Горбатов А.В., Налимов К.А., Абрамян М.А., Борисков М.В., Ковалев С.А., Свободов А.А., Архипов А.Н., Кривошеков Е.В., Амансахатова Е.Н., Тарасюк Е.С. Хирургическое лечение врожденных пороков сердца в Российской Федерации: данные национального регистра за 2024 год. *Патология кровообращения и кардиохирургия*. 2026;30(1):5-25. <https://doi.org/10.21688/1681-3472-2026-1-5-25>

Финансирование

Исследование не имело спонсорской поддержки.

Конфликт интересов

Авторы заявляют об отсутствии конфликта интересов.

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Аннотация

Актуальность. Врожденные пороки сердца остаются одной из ведущих причин младенческой смертности и инвалидизации, что обуславливает их высокую медико-социальную значимость. В условиях динамичного развития технологий, изменения демографических показателей и совершенствования пренатальной диагностики системный мониторинг результатов лечения на государственном уровне становится критически важным. Национальный регистр позволяет не только объективно оценить текущую эффективность кардиохирургической помощи в масштабах страны, но и выявить региональные тренды, а также определить приоритетные направления для внедрения современных гибридных и эндоваскулярных протоколов.

Цель. Оценить состояние и результаты хирургического лечения врожденных пороков сердца в Российской Федерации на основе данных национального мультицентрового регистра за 2024 г.

Методы. Проведен ретроспективный анализ 14 640 операций, выполненных в 31 кардиохирургическом центре. Для стратификации сложности вмешательства и оценки риска использовалась шкала RACHS II. Анализировались показатели госпитальной летальности, структура патологии и эффективность различных хирургических стратегий.

Результаты. Общая госпитальная летальность составила 1,42 %. В структуре вмешательств значительную долю (38,4 %) заняли эндоваскулярные процедуры. В сегменте интервенционной аритмологии ($n = 1342$) достигнута нулевая летальность. Наибольший риск сохраняется в группе новорожденных (летальность 6,6 %). Анализ паллиативной помощи выявил преимущество гибридного подхода: стентирование открытого артериального протока у дуктус-зависимых пациентов показало вдвое меньшую летальность (4,95 %) по сравнению с хирургическим системно-легочным анастомозом (9,74 %). Сложные реконструкции (процедура Norwood) остаются зоной высокого риска.

Заключение. Национальный регистр по детской кардиохирургии демонстрирует высокую эффективность и безопасность рутинных вмешательств. Выявлен отчетливый тренд на замещение открытой хирургии эндоваскулярными методами. Ключевыми факторами улучшения исходов при критических врожденных пороках сердца являются централизация помощи в экспертных центрах и внедрение гибридных технологий.

Ключевые слова: врожденные пороки сердца; госпитальная летальность; детская кардиохирургия; национальный регистр; эндоваскулярное лечение

Introduction

The contemporary epidemiological landscape of morbidity within the Russian Federation is undergoing substantial transformations, driven by a confluence of interrelated factors. Among the primary catalysts of these shifts are public and private sector investments in healthcare, which facilitate the modernization of medical infrastructure, the adoption of advanced diagnostic and therapeutic technologies, and enhanced accessibility of medical services [1]. Concurrently, the nation's economic progression, rising living standards, and evolving social structures exert a considerable influence on morbidity patterns. Equally critical macro-social determinants encompass demographic dynamics, including fluctua-

tions in birth rates and alterations in the frequency of pregnancy terminations. Recognized extensively within the scientific community, these factors constitute a multifaceted system impacting both individual and population-level health. Numerous scholars, including leading authorities in public health, underscore that social determinants of health – ranging from socioeconomic status and educational attainment to living conditions and access to adequate nutrition – exert a stratified and enduring influence on human well-being across the lifespan [2]. Consequently, comprehending and analyzing shifts in the disease spectrum necessitates a holistic approach that integrates not only biomedical factors but also the broader socioeconomic context.

Within the spectrum of contemporary pathology, congenital heart diseases (CHD) occupy a position of particular significance [3]. These complex conditions represent structural anomalies of the heart and great vessels arising during embryonic and fetal development. The etiology of CHD is multifactorial, involving intricate interactions between genetic predispositions and exposure to adverse environmental teratogens [3; 4]. According to numerous global epidemiological reports, the prevalence of CHD remains relatively stable, affecting approximately 0.8–1.0 % of all live births [5]. Notably, this frequency exhibits limited variability across different racial and ethnic groups, geographic regions, or nations with markedly disparate levels of economic development, underscoring the fundamental biological nature of these anomalies. However, a granular analysis of the specific spectrum of CHD phenotypes reveals regional variations. Select studies indicate a higher prevalence of left-sided cardiac lesions, such as coarctation of the aorta and hypoplastic left heart syndrome, within European populations [6–8]. Conversely, complex malformations like Tetralogy of Fallot are reportedly more frequent in Asian populations [9]. These findings suggest nuanced differences in CHD epidemiology, likely attributable to distinct genetic architectures among ethnic groups and variations in environmental exposures.

In the context of the Russian Federation, the Interregional Public Organization of Pediatric Cardiac Surgeons “Society of Specialists in Congenital Heart Diseases” conducts extensive research and ongoing surveillance, thereby generating unique data on regional characteristics of CHD. Observations derived from this organization’s activities largely corroborate global trends while simultaneously revealing patterns specific to the Russian population. Specifically, despite the overall stability in CHD prevalence, recent decades have witnessed significant shifts in the diagnostic profile [8; 9]. These transformations are attributable to a constellation of factors characteristic of contemporary Russian society. On one hand, substantial changes in the socioeconomic environment influence the accessibility and quality of medical care. On the other hand, the healthcare system is undergoing active development, with a pronounced emphasis on prenatal screening programs [5]. The implementation and refinement of advanced ultrasound diagnostic techniques in early pregnancy enable the detection of many congenital heart diseases (CHDs) prenatally, facilitating timely planning of perinatal management, delivery strategies, and subsequent surgical intervention [9]. Simultaneously, Russian society is experiencing considerable transformations in reproductive attitudes and behaviors, which inevitably impacts demographic structures

and, consequently, the overall epidemiology of congenital heart diseases. The cumulative effect of these factors is contributing to the emergence of a distinct spectrum of CHD in Russia, differentiating the country from other global regions not only in terms of aggregate statistics but also in the proportional distribution of specific nosological entities. These changes are manifested both in the dynamics of overall CHD case numbers and in the evolving profile of defect types.

The present study is dedicated to a comprehensive analysis and synthesis of extensive all-Russian data accumulated during the calendar year 2024. The data for this investigation were collated from 31 leading tertiary medical centers specializing in cardiac surgery, situated across diverse regions of the Russian Federation.

Methods

This investigation constitutes a retrospective analysis of a comprehensive database encompassing clinical and operative data from patients diagnosed with congenital heart diseases and cardiac arrhythmias. Data acquisition was conducted throughout the 2024 calendar year under the aegis of the Interregional Public Organization of Pediatric Cardiac Surgeons “Society of Specialists in Congenital Heart Diseases” and with the support of the Ministry of Health of the Russian Federation. This collaborative framework ensured a high degree of coordination, standardization of data collection protocols, and representativeness of the data at the national level.

Information was sourced from 31 major cardiac surgery centers operating within the Russian Federation. These institutions were selected based on their status as leading expert facilities, possessing substantial experience in the diagnosis and management of pediatric CHD and arrhythmias, as well as the technical infrastructure necessary for the standardized collection of demographic, clinical, diagnostic, and procedural data. Each participating center adhered strictly to unified data collection protocols, thereby ensuring data comparability and facilitating subsequent aggregation and analysis.

The study cohort comprised all patients who underwent surgical interventions for congenital heart diseases or cardiac arrhythmias during the reporting period (2024). Inclusion criteria were defined as the performance of any form of surgical or interventional procedure for the treatment of CHD, as well as surgical or interventional procedures for the correction of cardiac arrhythmias, irrespective of the patient’s age at the time of the procedure. The absence of a surgical intervention for the specified indications constituted the sole exclusion criterion.

To facilitate detailed analysis and stratified comparison of outcomes, all enrolled patients were categorized according to several key parameters. Age stratification was performed using the following groups: neonates (aged 0 to 28 days), infants (aged 29 days to 1 year), and older children (aged >1 year to 18 years). This stratification allowed for the consideration of age-specific pathophysiological characteristics and the inherent variability in surgical approaches across different developmental stages.

Based on the type of intervention performed, procedures were classified into the following categories: 1) open heart surgeries (requiring sternotomy or thoracotomy); 2) endovascular or interventional procedures (percutaneous catheter-based interventions); and 3) operations specifically aimed at the surgical or interventional correction of cardiac arrhythmias (e.g., pacemaker implantation, ablation procedures).

Further procedural details were documented according to the following parameters:

Procedure Primacy: Interventions were classified as primary if they constituted the first surgical procedure for the given CHD or arrhythmia, or as repeat if the patient had undergone previous surgical repair related to the primary diagnosis.

Procedure Intent: Interventions were categorized as radical (corrective) when aimed at complete anatomical and physiological restoration of normal cardiac function, or as palliative when intended to improve the patient's clinical status, alleviate symptoms, or serve as a preparatory stage for subsequent definitive correction.

Utilization of Cardiopulmonary Bypass (CPB): Procedures were stratified based on whether they were performed with or without the use of cardiopulmonary bypass, serving as a proxy for procedural complexity and invasiveness.

All aggregated data from the participating centers were centralized and stored within a secure, password-protected electronic data repository maintained at the Federal Center for Cardiovascular Surgery (Krasnoyarsk). This centralized approach ensured patient confidentiality, data integrity, facilitated efficient access for analytical purposes, and enabled rigorous quality control of the submitted data. Access to the database was strictly regulated and granted solely to authorized investigators for the explicit purposes of this research project.

The severity of surgical interventions was assessed using the RACHS-II (Risk Adjustment for Congenital Heart Surgery) consensus-based scoring system [10].

Operative mortality was defined as death occurring during the index hospitalization (irrespective of duration)

or within 30 days following the surgical procedure, if discharged.

Results

Overall Data

During the study period (2024), a total of 14,640 surgical interventions for congenital heart defects and cardiac arrhythmias were performed within the framework of 31 cardiac surgery centers across the Russian Federation. The overall postoperative mortality rate was 1.42 % (209 fatal outcomes).

The number of operations performed by each cardiac surgery center is presented in Table 1.

Patient Age Distribution at the Time of Surgery

Analysis of the age structure revealed that the predominant proportion consisted of children older than one year, accounting for 9,486 operations (64.8 %). This group exhibited the lowest mortality rate at 0.3 %. Children under one year of age (including neonates) underwent 5,154 operations (35.2 % of the total interventions), with a significantly higher mortality rate of 3.51 % in this cohort. Particular attention should be paid to the neonatal group (0–28 days of life), who underwent 1,542 operations (10.5 % of the total interventions). In this most vulnerable patient category, mortality reached 6.6 %. Within the neonatal group, low birth weight infants (weighing less than 2.5 kg) were specifically identified – 262 operations, constituting 17 % of all neonatal surgeries. Mortality in this subgroup was comparable to the overall neonatal mortality at 6.5 %. Among infants (aged 29 days to 1 year, excluding neonates), 3,612 operations were performed (24.7 % of the total) with a mortality rate of 2.1 %.

Types of Operations Performed

The structure of surgical interventions demonstrates a predominance of open heart surgeries, which accounted for 7,679 interventions (52.5 % of the total). Open surgeries were also characterized by the highest mortality rate at 2.5 % (190 fatal outcomes). Endovascular interventions comprised 5,619 operations (38.4 %), with a substantially lower mortality rate of 0.3 % (15 fatal outcomes). Operations for cardiac arrhythmias constituted the smallest proportion – 1,342 interventions (9.2 %) – and notably, no fatalities were registered in this category (0.0 %).

Characteristics of Surgical Interventions

The majority of performed procedures were primary interventions, totaling 9,012 operations (61.6 %), while repeat operations accounted for 1,987 cases (13.6 %). It is important to note that the sum of percentages for prima-

Table 1. Volume of Cardiac Surgery Operations in Various Cardiac Surgery Centers

Cardiac Surgery Center	Number of Operations
A.N. Bakulev National Medical Research Center for Cardiovascular Surgery, Moscow	2,709
E.N. Meshalkin National Medical Research Center, Novosibirsk	1,356
Federal Center for Cardiovascular Surgery, Astrakhan	829
Federal Center for Cardiovascular Surgery, Penza	760
V.A. Almazov National Medical Research Center, Saint Petersburg	698
Research Institute – S.V. Ochapovsky Regional Clinical Hospital No. 1, Krasnodar	621
Cardiology Research Institute, Tomsk National Research Medical Center, Tomsk	601
Morozov Children’s City Clinical Hospital, Moscow	556
S.G. Sukhanov Federal Center for Cardiovascular Surgery, Perm	531
Sverdlovsk Regional Clinical Hospital No. 1, Yekaterinburg	529
Federal Center for Cardiovascular Surgery, Krasnoyarsk	527
Children’s City Hospital No. 1, Saint Petersburg	506
Federal Center for Cardiovascular Surgery, Chelyabinsk	488
N.F. Filatov Children’s City Clinical Hospital No. 13, Moscow	402
Children’s Republican Clinical Hospital, Kazan	397
Federal Center for Cardiovascular Surgery, Khabarovsk	374
Irkutsk Order of the Badge of Honour Regional Clinical Hospital, Irkutsk	356
Regional Clinical Hospital No. 1, Tyumen	353
Republican Cardiology Center, Ufa	352
Research Institute for Complex Issues of Cardiovascular Diseases, Kemerovo	311
Federal Center for High Medical Technologies, Kaliningrad	262
Regional Children’s Clinical Hospital, Rostov-on-Don	204
V.P. Polyakov Samara Regional Clinical Cardiology Dispensary, Samara	194
District Cardiology Dispensary "Center for Diagnostics and Cardiovascular Surgery", Surgut	172
Volgograd Regional Clinical Cardiology Center, Volgograd	117
B.A. Korolev Research Institute – Specialized Cardiac Surgery Clinical Hospital, Nizhny Novgorod	109
L.M. Roshal Children’s Clinical Center, Krasnogorsk	96
Voronezh Regional Clinical Hospital No. 1, Voronezh	95
M.E. Nikolaev Republican Hospital No. 1 – National Medicine Center, Yakutsk	67
Regional Clinical Hospital, Omsk	34
Irkutsk State Regional Children’s Clinical Hospital, Irkutsk	34

ry and repeat operations does not reach 100 %, indicating the possible absence of this gradation for a portion of the interventions.

Regarding the intent of the intervention, the following distribution was observed: curative surgeries (aimed at complete defect repair) constituted 6,289 cases (43 %). Palliative interventions (aimed at improving the patient’s condition prior to potential radical correction or in cases where the defect is inoperable) occupied a significantly

smaller proportion – 229 operations (1.6 %). Thus, a considerably larger portion of operations was directed towards the complete repair of the existing CHD.

Operations with Cardiopulmonary Bypass: The use of cardiopulmonary bypass is an important indicator of the complexity and extent of the operation. A total of 6,025 operations (41.2 % of the total) were performed using CPB. In this group, the overall mortality rate was 2.4 % (145 fatal outcomes).

A more detailed analysis of mortality in the CPB groups revealed the following trends. CPB operations in neonates: among 638 CPB interventions performed on neonates, mortality was 9.2 % (59 fatal outcomes). This represents the highest mortality rate among all CPB and age subgroups, attributable to the extreme vulnerability of this age category, low body weight, and the complexity of the pathologies. CPB operations in children under 1 year: in this group (including neonates), 3,019 operations were performed using CPB with a mortality rate of 3.9% (119 fatal outcomes). CPB operations in children older than 1 year: 3,006 operations using CPB were performed on children older than one year. This group recorded the lowest mortality among CPB operations – 0.9 % (26 fatal outcomes).

In 2024, 9,012 new cases of congenital heart diseases were diagnosed. The types of diagnosed CHDs are presented in Table 2.

As can be seen from the data in Table 2, the most frequent CHDs were atrial septal defect – 24.77 %; patent ductus arteriosus – 18.13 %; ventricular septal defect – 16.63 %; coarctation of the aorta – 7.07 %; valvular pulmonary stenosis – 5.13 %; Tetralogy of Fallot – 4.41 %; transposition of the great arteries – 3.27 %; aortic valve stenosis – 2.94 %; atrioventricular septal defect – 2.81 %; and partial anomalous pulmonary venous return – 2.66 % (Fig. 1).

Risk Stratification According to the RACHS-II Consensus Scoring System

All surgical procedures were stratified according to the RACHS-II (Risk Adjustment for Congenital Heart Surgery) consensus-based scoring system. The distribution of Category 1 risk stratification is presented in Table 3.

As demonstrated in Table 3, the total number of surgical interventions classified as RACHS-II Category 1 was 8,044 procedures, with an associated mortality rate of 0.33 % (27 patients).

The distribution of Category 2 risk stratification is presented in Table 4.

As demonstrated in Table 4, the total number of surgical interventions classified as RACHS-II Category 2 was 4,858 procedures, with an associated mortality rate of 0.98 % (48 patients).

The distribution of Category 3 risk stratification is presented in Table 5.

As demonstrated in Table 5, the total number of surgical interventions classified as RACHS-II Category 3 was 587 procedures, with an associated mortality rate of 6.47 % (38 patients).

The distribution of Category 4 risk stratification is presented in Table 6.

Table 2. Types of Congenital Heart Defects Diagnosed in 2024

Congenital Heart Defect	Number of Patients	Proportion of Total CHD, %
ASD	2,232	24.78
VSD	1,499	16.63
AVSD	253	2.81
AP Window	18	0.20
Anomalous Origin of Pulmonary Artery from Aorta	5	0.06
Truncus Arteriosus	26	0.29
PAPVR	240	2.66
TAPVR	91	1.01
Cor Triatriatum	31	0.34
Anomalies of Systemic Venous Return	3	0.03
Tetralogy of Fallot	397	4.41
Pulmonary Atresia with VSD	127	1.41
Tricuspid Valve Pathology	60	0.67
Pulmonary Atresia with Intact Ventricular Septum	37	0.41
Valvular Pulmonary Stenosis	462	5.13
Aortic Valve Stenosis	265	2.94
Aortic Root Aneurysm	12	0.13
LVOT Obstruction	80	0.89
Supravalvular Aortic Stenosis	19	0.21
Aorto-Left Ventricular Tunnel	10	0.11
Mitral Valve Pathology	75	0.83
Hypoplastic Left Heart Syndrome	48	0.53
Single Ventricle	192	2.13
Transposition of the Great Arteries	295	3.27
Congenitally Corrected Transposition of the Great Arteries	11	0.12
Coarctation of the Aorta	637	7.07
Interrupted Aortic Arch	40	0.44
Coronary-Pulmonary Fistula	13	0.14
Anomalous Origin of Coronary Artery from Pulmonary Artery	21	0.23
Patent Ductus Arteriosus	1,634	18.13
Vascular Ring	97	1.08
DORV	82	0.91

Note. AP Window – aortopulmonary window; ASD – atrial septal defect; AVSD – atrioventricular septal defect; DORV – double outlet right ventricle; LVOT – left ventricular outflow tract; PAPVR – partial anomalous pulmonary venous return; TAPVR – total anomalous pulmonary venous return; VSD – ventricular septal defect.

Fig. 1. Most Prevalent Congenital Heart Defect Phenotypes (accounting for 87.82 % of the total congenital heart disease)

Note. AoS – aortic valve stenosis; ASD – atrial septal defect; AVSD – atrioventricular septal defect; CoA – coarctation of the aorta; PAPVR – partial anomalous pulmonary venous return; PDA – patent ductus arteriosus; TGA – transposition of the great arteries; TOF – Tetralogy of Fallot; VPS – valvular pulmonary stenosis; VSD – ventricular septal defect.

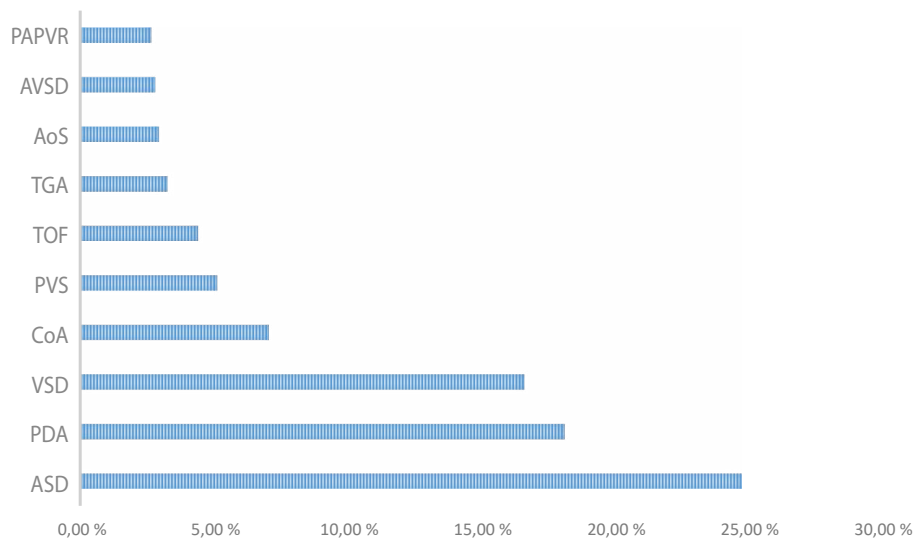


Table 3. RACHS-II Category 1 Risk Stratification for Congenital Heart Surgery

Procedure	Procedural Volume	Mortality, %
ASD Closure (Surgical)	1,526	0.06
VSD Closure (Surgical)	1,322	0.45
Correction of Partial AVSD	131	1.5
Correction of PAPVR	246	0.40
Correction of Cor Triatriatum	31	0
Closure of MAPCAs	351	0
Tricuspid Valve Reconstruction	83	1.20
Pulmonary Artery Trunk Reconstruction	42	0
Pulmonary Valve Commissurotomy	81	1.23
Pulmonary Valve Replacement	154	0.64
Aortic Valve Plastic Repair	176	0
Aortic Valve Replacement	130	0
Ozaki Procedure (Aortic Valve Neocuspidization)	26	3.84
Relief of LVOT Obstruction	28	3.57
Resection of Fibro-muscular Membrane (Subvalvular)	48	0
Extended LVOT Myectomy	38	5.26
Correction of Supravalvular Aortic Stenosis	24	4.16
Correction of Aorto-Left Ventricular Tunnel	12	0
VSD Closure in CCTGA	4	0
Correction of Coarctation of the Aorta	491	1.01
Resection of Coarctation with Aortic Prosthetic Replacement	20	5.00
Vascular Ring Division	183	0.54
Aortic Valve Balloon Angioplasty	68	0
Pulmonary Valve Balloon Angioplasty	335	0.59
ASD Device Closure	1,007	0
VSD Device Closure	94	0
Closure of Veno-venous Collaterals	46	0
Closure of Previously Created Fenestrations	5	0
Arrhythmia Surgery (All Types)	1,342	0

Note. ASD – atrial septal defect; AVSD – atrioventricular septal defect; CCTGA – congenitally corrected transposition of the great arteries; LVOT – left ventricular outflow tract; MAPCAs – major aortopulmonary collateral arteries; PAPVR – partial anomalous pulmonary venous return; VSD – ventricular septal defect.

Table 4. RACHS-II Category 2 Risk Stratification for Congenital Heart Surgery

Procedure	Procedural Volume	Mortality, %
Correction of Complete AVSD	181	2.20
Correction of TOF with Pulmonary Valve Annulus Preservation	166	0.60
Correction of TOF with Transannular Patch	159	4.40
Correction of TOF with RV-PA Conduit Implantation	19	0
Correction of RVOT Stenosis	107	1.86
Resection of RVOT Aneurysm	3	0
Reconstruction of Pulmonary Artery Branches	48	2.08
Implantation of RV-PA Conduit	175	1.71
Mitral Valve Plastic Repair	63	1.58
Resection of Supramitral Ring	5	0
Fontan Operation	50	4.00
Arterial Switch Operation (ASO)	177	7.90
Senning Operation	1	0
Resection of Coarctation and VSD Closure	44	11.36
Closure of Coronary Fistulae	20	0
Correction of Anomalous Origin of Coronary Artery from Pulmonary Artery (ALCAPA)	26	0
PDA Closure (Surgical)	1,322	0.07
Creation of Bidirectional Cavopulmonary Anastomosis (BCPA)	104	3.84
Hemi-Fontan Operation	1	0
PDA Device or Coil Closure	1,581	0.06
Balloon Angioplasty of Pulmonary Artery Branches	306	0
Stenting of Pulmonary Artery Branches	57	1.75
Balloon Angioplasty or Stenting of Great Vessels	235	0.42
Stenting of Systemic Veins	8	0

Note. ALCAPA – anomalous left coronary artery from pulmonary artery; AVSD – atrioventricular septal defect; PDA – patent ductus arteriosus; RVOT – right ventricular outflow tract; RV-PA – right ventricle to pulmonary artery; TOF – Tetralogy of Fallot; VSD – ventricular septal defect.

Table 5. RACHS-II Category 3 Risk Stratification for Congenital Heart Surgery

Procedure	Procedural Volume	Mortality, %
Creation of LV-Aorta Tunnel	11	0
Radical Correction of Pulmonary Atresia with VSD	74	12.16
Unifocalization of Pulmonary Arteries	74	6.75
One-and-a-Half Ventricle Repair	4	0
Ross Procedure (Pulmonary Autograft)	32	3.12
Konno Procedure (Aortic Ventriculoplasty)	12	0
Mitral Valve Replacement	45	0
Arterial Switch Operation and VSD Closure	64	9.37
Mustard Operation with VSD Closure	1	0
Radical Correction of TGA with VSD and LVOT Stenosis	3	0
Rastelli Operation (RV-PA Conduit with VSD Closure)	7	0
Yamagishi Procedure (Modified Blalock-Taussig Shunt)	1	0
Double Switch Operation (Atrial + Arterial Switch)	8	12.5
Atrioventricular Valve Replacement in CCTGA	4	0
Aortoplasty for Interrupted Aortic Arch	46	8.69
Rashkind Procedure (Balloon Atrial Septostomy)	118	3.38
Stenting of ASD	19	15.78
Aortic Root Replacement	32	9.37
Mitral Valve Replacement	11	0
Aortic Valve Replacement	15	13.3
Tricuspid Valve Replacement	3	0
Resection of Cardiac Tumors	3	0

Note. ASD – atrial septal defect; AV – atrioventricular; CCTGA – congenitally corrected transposition of the great arteries; LV – left ventricle; LVOT – left ventricular outflow tract; PA – pulmonary atresia; RV-PA – right ventricle to pulmonary artery; TGA – transposition of the great arteries; VSD – ventricular septal defect.

As demonstrated in Table 6, the total number of surgical interventions classified as RACHS-II Category 4 was 1,046 procedures, with an associated mortality rate of 7.17 % (75 patients).

The distribution of Category 5 risk stratification is presented in Table 7.

As demonstrated in Table 7, the total number of surgical interventions classified as RACHS-II Category 5 was 65 procedures, with an associated mortality rate of 24.61 % (16 patients).

RACHS-II Risk Stratification in the Neonatal Cohort (Age < 28 Days)

Among neonates, 1,542 surgical interventions were performed, yielding an aggregate mortality rate of 6.6 % (102 patients). Of these, 638 procedures (constituting 41.4 % of the neonatal cohort) were performed utilizing cardiopulmonary bypass, with an associated mortality rate of 9.2 % in this subgroup.

The distribution of RACHS-II risk categories within the neonatal cohort is presented in Tables 8–12.

As demonstrated in Table 8, the total number of neonatal surgical interventions classified as RACHS-II Category 1 was 459 procedures, with an associated mortality rate of 2.39 % (11 patients).

As demonstrated in Table 9, the total number of neonatal surgical interventions classified as RACHS-II Category 2 was 329 procedures, with an associated mortality rate of 4.86 % (16 patients).

As demonstrated in Table 10, the total number of neonatal surgical interventions classified as RACHS-II Category 3 was 233 procedures, with an associated mortality rate of 9.01 % (21 patients).

As demonstrated in Table 11, the total number of neonatal surgical interventions classified as RACHS-II Category 4 was 481 procedures, with an associated mortality rate of 9.14 % (44 patients).

As demonstrated in Table 12, the total number of neonatal surgical interventions classified as RACHS-II Category 5 was 40 procedures, with an associated mortality rate of 25 % (10 patients).

A graphical representation of RACHS-II risk stratification distribution within the neonatal cohort is presented in Fig. 2.

RACHS-II Risk Stratification for Surgical Treatment in Infants (Aged 29 Days to 12 Months)

Among infants, a total of 3,612 surgical interventions were performed, yielding an aggregate mortality rate of 2.18 % (79 patients). Of these, 2,381 procedures (constituting 65.9 % of the infant cohort) were performed utilizing cardiopulmonary bypass, with an associated mortality rate of 3.9 % in this subgroup.

Table 6. RACHS-II Category 4 Risk Stratification for Congenital Heart Surgery

Procedure	Procedural Volume	Mortality, %
Correction of Truncus Arteriosus	32	6.25
Correction of Truncus Arteriosus with Interrupted Aortic Arch	6	16.6
Correction of TAPVR	94	8.51
Correction of Pulmonary Vein Stenosis	10	10
Correction of Anomalies of Venous Return	4	0
Stenting of RVOT	29	6.89
Take-down of Fontan Circulation	1	0
Resection of Coarctation and Pulmonary Artery Banding	112	5.35
Stenting of PDA	101	4.95
Creation of Systemic-to-Pulmonary Artery Shunt	154	9.74
Creation of Direct Aorta-to-Pulmonary Artery Anastomosis	21	19.04
Creation of RV-PA Shunt	6	0
Closure of Systemic-to-Pulmonary Artery Shunt	3	0
Pulmonary Artery Banding (PAB)	330	3.93
Removal of Pulmonary Artery Band	75	1.33
Banding of Pulmonary Artery Branches and PDA Stenting	68	25

Note. PDA – patent ductus arteriosus; RVOT – right ventricular outflow tract; RV-PA – right ventricle to pulmonary artery; TA – truncus arteriosus; TAPVR – total anomalous pulmonary venous return.

Table 7. RACHS-II Category 5 Risk Stratification for Congenital Heart Surgery

Procedure	Procedural Volume	Mortality, %
Norwood Procedure (Stage I Palliation for HLHS)	47	29.78
Yasui Operation	2	0
Damus – Kaye – Stansel Operation	16	12.5

Note. HLHS – hypoplastic left heart syndrome; IAA – interrupted aortic arch; LVOT – left ventricular outflow tract; VSD – ventricular septal defect.

Table 8. RACHS-II Category 1 Risk Stratification for Congenital Heart Surgery (Neonatal Cohort)

Procedure	Procedural Volume	Mortality, %
Correction of Coarctation of the Aorta	234	2.1
Pulmonary Valve Balloon Angioplasty	69	2.9
Aortic Valve Balloon Angioplasty	38	0
Aortic Valve Plastic Repair	34	0
Vascular Ring Division	19	0
VSD Closure	18	5.6
Pulmonary Valve Commissurotomy	14	7.10
Pulmonary Artery Trunk Reconstruction	10	0
Epicardial Permanent Pacemaker Implantation	5	0
Resection of Coarctation with Aortic Prosthetic Replacement	3	33.3
Correction of PAPVR	2	50
Tricuspid Valve Reconstruction	4	0
Relief of LVOT Obstruction	2	0
ASD Closure	1	0
Pulmonary Valve Replacement	1	0
Ozaki Procedure	1	0
Resection of Fibro-muscular Membrane	1	0
Correction of Aorto-Left Ventricular Tunnel	1	0
Closure of MAPCAs	2	0

Note. ASD – atrial septal defect; LVOT – left ventricular outflow tract; MAPCAs – major aortopulmonary collateral arteries; PAPVR – partial anomalous pulmonary venous return; VSD – ventricular septal defect.

All infant procedures were stratified according to the RACHS-II consensus scoring system. The distribution of Category 1 risk stratification is presented in Table 13.

As demonstrated in Table 13, the total number of infant surgical interventions classified as RACHS-II Category 1 was 1,977 procedures, with an associated mortality rate of 0.60 % (12 patients).

The distribution of Category 2 risk stratification in the infant cohort is presented in Table 14.

As demonstrated in Table 14, the total number of infant surgical interventions classified as RACHS-II Category 2 was 1,014 procedures, with an associated mortality rate of 2.17 % (22 patients).

The distribution of Category 3 risk stratification in the infant cohort is presented in Table 15.

Table 9. RACHS-II Category 2 Risk Stratification for Congenital Heart Surgery (Neonatal Cohort)

Procedure	Procedural Volume	Mortality, %
Arterial Switch Operation (ASO)	155	7.1
PDA Closure (Surgical)	101	0
Resection of Coarctation with VSD Closure	25	16
Balloon Angioplasty or Stenting of Great Vessels	11	9.1
Correction of RVOT Stenosis	10	0
Correction of TOF	11	0
Reconstruction of Pulmonary Artery Branches	6	0
Correction of Complete AVSD	2	0
Endovascular PDA Closure	3	0
Balloon Angioplasty of Pulmonary Artery Branches	2	0
Closure of Coronary Artery Fistula	1	0
Implantation of RV-PA Conduit	2	0

Note. AVSD – atrioventricular septal defect; PDA – patent ductus arteriosus; RVOT – right ventricular outflow tract; RV-PA – right ventricle to pulmonary artery; TOF – Tetralogy of Fallot; VSD – ventricular septal defect.

Table 10. RACHS-II Category 3 Risk Stratification for Congenital Heart Surgery (Neonatal Cohort)

Procedure	Procedural Volume	Mortality, %
Rashkind Procedure (Balloon Atrial Septostomy)	111	3.6
Arterial Switch Operation and VSD Closure	50	12
Aortoplasty for Interrupted Aortic Arch	36	11.1
Unifocalization of Pulmonary Arteries	17	11.8
Stenting of ASD	5	60
Radical Correction of Pulmonary Atresia with VSD	8	25
Konno Procedure	4	0
Correction of DORV from RV	1	0
Double Switch Operation	1	0

Note. ASD – atrial septal defect; DORV from RV – double outlet right ventricle; VSD – ventricular septal defect.

Table 11. RACHS-II Category 4 Risk Stratification for Congenital Heart Surgery (Neonatal Cohort)

Procedure	Procedural Volume	Mortality, %
Radical Correction of Truncus Arteriosus	10	0
Correction of Truncus Arteriosus with Interrupted Aortic Arch	2	0
Correction of TAPVR	64	9.4
Correction of Pulmonary Vein Stenosis	1	0
Stenting of RVOT	15	13.3
Resection of Coarctation and Pulmonary Artery Banding	80	5
Stenting of PDA	53	7.5
Creation of Systemic-to-Pulmonary Artery Shunt	74	10.8
Creation of Direct Aorta-to-Pulmonary Artery Anastomosis	6	16.7
Creation of RV-PA Shunt	1	0
Closure of Systemic-to-Pulmonary Artery Shunt	1	0
Pulmonary Artery Banding (PAB)	115	6.1
Banding of Pulmonary Artery Branches and PDA Stenting	59	20.3

Note. PDA – patent ductus arteriosus; RVOT – right ventricular outflow tract; RV-PA – right ventricle to pulmonary artery; TAPVR – total anomalous pulmonary venous return.

Table 12. RACHS-II Category 5 Risk Stratification for Congenital Heart Surgery (Neonatal Cohort)

Procedure	Procedural Volume	Mortality, %
Norwood Procedure (Stage I Palliation for HLHS)	31	32.3
Yasui Operation	2	0
Damus – Kaye – Stansel Operation	7	0

Note. HLHS – hypoplastic left heart syndrome.

As demonstrated in Table 15, the total number of infant surgical interventions classified as RACHS-II Category 3 was 125 procedures, with an associated mortality rate of 7.20 % (9 patients).

The distribution of Category 4 risk stratification in the infant cohort is presented in Table 16.

As demonstrated in Table 16, the total number of infant surgical interventions classified as RACHS-II Category 4 was 474 procedures, with an associated mortality rate of 6.32 % (30 patients).

The distribution of Category 5 risk stratification in the infant cohort is presented in Table 17.

As demonstrated in Table 17, the total number of infant surgical interventions classified as RACHS-II Category 5 was 22 procedures, with an associated mortality rate of 27.27 % (6 patients).

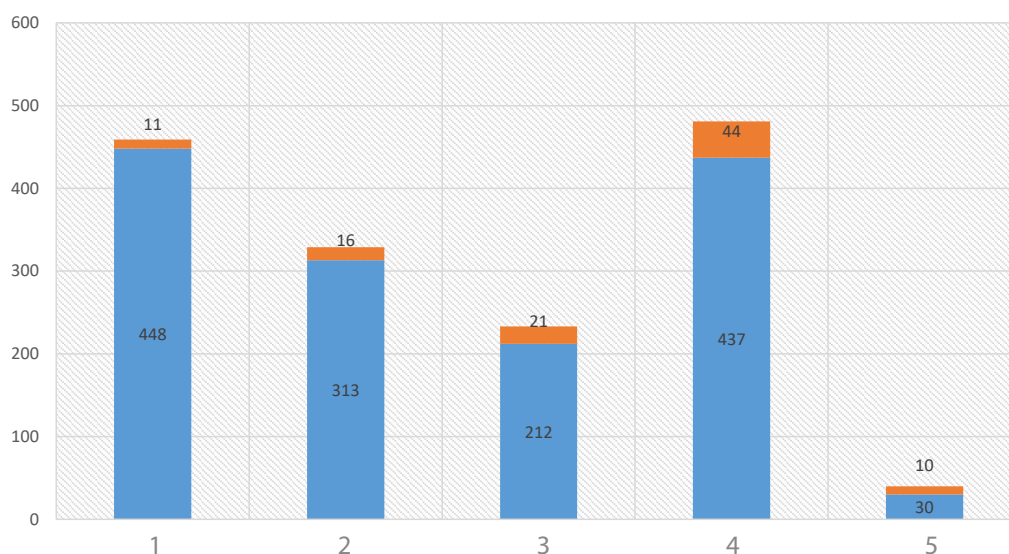


Fig. 2. RACHS-II Risk Stratification for Surgical Treatment in Neonates. Orange indicates deceased patients, blue indicates discharged patients, and the numbers represent the RACHS-II risk category

Table 13. RACHS-II Category 1 Risk Stratification for Congenital Heart Surgery (Infant Cohort)

Procedure	Procedural Volume	Mortality, %
ASD Closure (Surgical)	237	0
VSD Closure (Surgical)	901	0.60
Correction of Partial AVSD	43	4.65
Correction of PAPVR	31	0
Correction of Cor Triatriatum	11	0
Closure of MAPCAs	52	0
Tricuspid Valve Reconstruction	4	25
Pulmonary Artery Trunk Reconstruction	32	0
Pulmonary Valve Commissurotomy	40	0
Pulmonary Valve Replacement	30	0
Aortic Valve Plastic Repair	57	0
Ozaki Procedure	2	0
Relief of LVOT Obstruction	26	3.84
Resection of Fibro-muscular Membrane	5	0
Extended LVOT Myectomy	16	12.5
Correction of Supravalvular Aortic Stenosis	4	0
Correction of Aorto-Left Ventricular Tunnel	5	0
Correction of Coarctation of the Aorta	169	0
Resection of Coarctation with Aortic Prosthetic Replacement	2	0
Vascular Ring Division	107	0.93
Aortic Valve Balloon Angioplasty	22	0
Pulmonary Valve Balloon Angioplasty	140	0
ASD Device Closure	4	0
VSD Device Closure	8	0
Closure of Venovenous Collaterals	4	0
Epicardial Pacemaker Implantation	21	0
Implantation of Cardioverter-Defibrillator	4	0

Note. ASD – atrial septal defect; AVSD – atrioventricular septal defect; LVOT – left ventricular outflow tract; MAPCAs – major aortopulmonary collateral arteries; PAPVR – partial anomalous pulmonary venous return; VSD – ventricular septal defect.

Table 14. RACHS-II Category 2 Risk Stratification for Congenital Heart Surgery (Infant Cohort)

Procedure	Procedural Volume	Mortality, %
Correction of Complete AVSD	145	2.75
Correction of TOF with Pulmonary Valve Annulus Preservation	133	0.75
Correction of TOF with Transannular Patch	138	3.62
Correction of TOF with RV-PA Conduit Implantation	12	0
Correction of RVOT Stenosis	43	2.32
Resection of RVOT Aneurysm	2	0
Reconstruction of Pulmonary Artery Branches	14	7.14
Implantation of RV-PA Conduit	19	0
Mitral Valve Plastic Repair	2	0
Resection of Supramitral Ring	2	0
Arterial Switch Operation (ASO)	17	11.76
Senning Operation	1	0
Resection of Coarctation and VSD Closure	15	6.66
Closure of Coronary Fistulae	6	0
Correction of ALCAPA	16	0
PDA Closure (Surgical)	188	0.53
Creation of Bidirectional Cavopulmonary Anastomosis (BCPA)	70	5.71
Hemi-Fontan Operation	1	0
PDA Device or Coil Closure	79	1.26
Balloon Angioplasty of Pulmonary Artery Branches	40	0
Stenting of Pulmonary Artery Branches	7	14.28
Balloon Angioplasty or Stenting of Great Vessels	63	0
Stenting of Systemic Veins	1	0

Note. ALCAPA – anomalous left coronary artery from pulmonary artery; AVSD – atrioventricular septal defect; PDA – patent ductus arteriosus; RVOT – right ventricular outflow tract; RV-PA – right ventricle to pulmonary artery; TOF – Tetralogy of Fallot; VSD – ventricular septal defect.

Table 15. RACHS-II Category 3 Risk Stratification for Congenital Heart Surgery (Infant Cohort)

Procedure	Procedural Volume	Mortality, %
Correction of Pulmonary Atresia with VSD	36	16.66
Unifocalization of Pulmonary Arteries	30	3.33
Aortoplasty for Interrupted Aortic Arch	10	0
One-and-a-Half Ventricle Repair	1	0
Ross Procedure	4	25
Konno Procedure	4	0
Mitral Valve Replacement	5	0
Arterial Switch Operation and VSD Closure	13	0
Correction of TGA with VSD and LVOT Stenosis	1	0
Rastelli Operation	1	0
Double Switch Operation	2	50
Rashkind Procedure	7	0
Stenting of ASD	6	0
Aortic Root Replacement	2	0
Resection of Cardiac Tumors	3	0

Note. ASD – atrial septal defect; LVOT – left ventricular outflow tract; TGA – transposition of the great arteries; VSD – ventricular septal defect.

RACHS-II Risk Stratification for Surgical Treatment in Children Older Than One Year (Aged 1 Year to 18 Years)

Among patients aged 1 year to 18 years, a total of 9,486 surgical interventions were performed, yielding an aggregate mortality rate of 0.29 % (28 patients). Of these, 3,019 procedures (constituting 31.8 % of this cohort) were performed utilizing cardiopulmonary bypass, with an associated mortality rate of 0.9 % in this subgroup.

All procedures in patients aged 1 to 18 years were stratified according to the RACHS-II consensus scoring system. The distribution of Category 1 risk stratification is presented in Table 18.

As demonstrated in Table 18, the total number of surgical interventions in patients aged 1–18 years classified as RACHS-II Category 1 was 5,612 procedures, with an associated mortality rate of 0.07 % (4 patients).

The distribution of Category 2 risk stratification in the cohort aged 1–18 years is presented in Table 19.

Table 16. RACHS-II Category 4 Risk Stratification for Congenital Heart Surgery (Infant Cohort)

Procedure	Procedural Volume	Mortality, %
Correction of Truncus Arteriosus	13	15.38
Correction of Truncus Arteriosus with Interrupted Aortic Arch	4	25
Correction of TAPVR	28	7.14
Correction of Pulmonary Vein Stenosis	7	14.28
Correction of Anomalies of Venous Return	1	0
Stenting of RVOT	14	0
Resection of Coarctation and Pulmonary Artery Banding	30	6.66
Stenting of PDA	28	0
Creation of Systemic-to-Pulmonary Artery Shunt	64	10.93
Creation of Direct Aorta-to-Pulmonary Artery Anastomosis	15	20
Creation of RV-PA Shunt	1	0
Closure of Systemic-to-Pulmonary Artery Shunt	2	0
Pulmonary Artery Banding (PAB)	203	2.95
Removal of Pulmonary Artery Band	55	1.81
Banding of Pulmonary Artery Branches and PDA Stenting	9	55.6

Note. PDA – patent ductus arteriosus; RVOT – right ventricular outflow tract; RV-PA – right ventricle to pulmonary artery; TA – truncus arteriosus; TAPVR – total anomalous pulmonary venous return.

As demonstrated in Table 19, the total number of surgical interventions in patients aged 1–18 years classified as RACHS-II Category 2 was 3,550 procedures, with an associated mortality rate of 0.28 % (10 patients).

The distribution of Category 3 risk stratification in the cohort aged 1–18 years is presented in Table 20.

As demonstrated in Table 20, the total number of surgical interventions in patients aged 1–18 years classified as RACHS-II Category 3 was 229 procedures, with an associated mortality rate of 3.49 % (8 patients).

The distribution of Category 4 risk stratification in the cohort aged 1–18 years is presented in Table 21.

As demonstrated in Table 21, the total number of surgical interventions in patients aged 1–18 years classified as RACHS-II Category 4 was 91 procedures, with an associated mortality rate of 1.09 % (1 patient).

Table 17. RACHS-II Category 5 Risk Stratification for Congenital Heart Surgery (Infant Cohort)

Procedure	Procedural Volume	Mortality, %
Norwood Procedure	16	25
Damus – Kaye – Stansel Operation	6	33.3

The distribution of Category 5 risk stratification in the cohort aged 1–18 years is presented in Table 22.

As demonstrated in Table 22, the total number of surgical interventions in patients aged 1–18 years classified as RACHS-II Category 5 was 3 procedures, with no associated fatalities.

Discussion

The present study, encompassing data from 31 leading cardiac surgery centers of the Russian Federation for the year 2024, represents one of the most extensive publications on the state of pediatric cardiac surgery in the country. The analysis of 14,640 surgical interventions permits not only an assessment of the current efficacy of surgical care but also the identification of systemic patterns shaping the development of the specialty. The overall hospital mortality rate of 1.42 % observed in the study cohort demonstrates a high level of safety in cardiac surgical care in Russia, comparable to data from leading international registries. For comparative purposes, data from The Society of Thoracic Surgeons (STS) and the European Association for Cardio-Thoracic Surgery (EACTS) databases in recent years show aggregated mortality rates in the range of 2.5–3.0 % for the entire population [11; 12]; however, direct comparison requires caution due to differences in patient case mix and complexity. Nevertheless, the achieved rate of 1.42 % attests to the maturity of the national school of pediatric cardiac surgery and the effectiveness of patient referral pathways.

Analysis of the distribution of nosological forms confirms global epidemiological patterns. The predominance of septal defects (atrial septal defect (ASD) – 24.77 %, ventricular septal defect (VSD) – 16.63 %) and patent ductus arteriosus (18.13 %) correlates with worldwide statistics, where these defects constitute up to 50 % of all congenital cardiac anomalies [4]. However, particular attention is drawn to the high detection rate and surgical activity concerning critical defects in the neonatal period. The proportions of coarctation of the aorta (7.07 %), Tetralogy of Fallot (4.41 %), and transposition of the great arteries (3.27 %) indicate the quality of the prenatal diagnostic and neonatal screening systems. In contemporary cardiac surgery, timely diagnosis is the key to reducing mortality. The fact that

Table 18. RACHS-II Category 1 Risk Stratification for Congenital Heart Surgery (Cohort Aged 1–18 Years)

Procedure	Procedural Volume	Mortality, %
ASD Closure (Surgical)	1,285	0.07
VSD Closure (Surgical)	403	0
Correction of Partial AVSD	88	0
Correction of PAPVR	213	0
Correction of Cor Triatriatum	20	0
Closure of MAPCAs	297	0
Tricuspid Valve Reconstruction	75	0
Pulmonary Valve Commissurotomy	27	0
Pulmonary Valve Replacement	123	0.81
Aortic Valve Plastic Repair	85	0
Aortic Valve Replacement	130	0
Ozaki Procedure	23	4.34
Resection of Fibro-muscular Membrane	42	0
Extended LVOT Myectomy	22	0
Correction of Supravalvular Aortic Stenosis	20	5
Correction of Aorto-Left Ventricular Tunnel	6	0
VSD Closure in CCTGA	4	0
Correction of Coarctation of the Aorta	88	0
Resection of Coarctation with Aortic Prosthetic Replacement	15	0
Vascular Ring Division	57	0
Aortic Valve Balloon Angioplasty	8	0
Pulmonary Valve Balloon Angioplasty	126	0
ASD Device Closure	1,003	0
VSD Device Closure	86	0
Closure of Venovenous Collaterals	45	0
Closure of Previously Created Fenestrations	5	0
Radiofrequency Ablation of Accessory Pathways	1,061	0
Endocardial Pacemaker Implantation	148	0
Epicardial Pacemaker Implantation	94	0
Implantation of Cardioverter-Defibrillator	11	0

Note. ASD – atrial septal defect; AVSD – atrioventricular septal defect; CCTGA – congenitally corrected transposition of the great arteries; LVOT – left ventricular outflow tract; MAPCAs – major aortopulmonary collateral arteries; PAPVR – partial anomalous pulmonary venous return; VSD – ventricular septal defect.

Table 19. RACHS-II Category 2 Risk Stratification for Congenital Heart Surgery (Cohort Aged 1–18 Years)

Procedure	Procedural Volume	Mortality, %
Correction of Complete AVSD	34	0
Correction of TOF with Pulmonary Valve Annulus Preservation	30	0
Correction of TOF with Transannular Patch	13	15.38
Correction of TOF with RV-PA Conduit Implantation	7	0
Correction of RVOT Stenosis	54	1.85
Resection of RVOT Aneurysm	1	0
Reconstruction of Pulmonary Artery Branches	28	0
Implantation of RV-PA Conduit	154	1.94
Mitral Valve Plastic Repair	61	1.63
Resection of Supramitral Ring	3	0
Fontan Operation	50	4.00
Arterial Switch Operation (ASO)	5	20
Resection of Coarctation and VSD Closure	4	0
Closure of Coronary Fistulae	13	0
Correction of ALCAPA	10	0
PDA Closure (Surgical)	1,033	0
Creation of Bidirectional Cavopulmonary Anastomosis (BCPA)	34	0
PDA Device or Coil Closure	1,544	0
Balloon Angioplasty of Pulmonary Artery Branches	264	0
Stenting of Pulmonary Artery Branches	50	0
Balloon Angioplasty or Stenting of Great Vessels	161	0
Stenting of Systemic Veins	7	0

Note. ALCAPA – anomalous left coronary artery from pulmonary artery; AVSD – atrioventricular septal defect; PDA – patent ductus arteriosus; RV-PA – right ventricle to pulmonary artery; TOF – Tetralogy of Fallot; RVOT – right ventricular outflow tract; VSD – ventricular septal defect.

Table 20. RACHS-II Category 3 Risk Stratification for Congenital Heart Surgery (Cohort Aged 1–18 Years)

Procedure	Procedural Volume	Mortality, %
Creation of LV-Aorta Tunnel	10	0
Correction of Pulmonary Atresia with VSD	20	5
Unifocalization of Pulmonary Arteries	37	5.40
One-and-a-Half Ventricle Repair	3	0
Ross Procedure	28	0
Konno Procedure	4	0
Mitral Valve Replacement	40	0
Arterial Switch Operation and VSD Closure	1	0
Mustard Operation with VSD Closure	1	0
Correction of TGA with VSD and LVOT Stenosis	2	0
Rastelli Operation	6	0
Yamagishi Procedure	1	0
Double Switch Operation	5	0
Atrioventricular Valve Replacement in CCTGA	4	0
Stenting of ASD	8	0
Aortic Root Replacement	30	10
Mitral Valve Re-replacement	11	0
Aortic Valve Re-replacement	15	13.3
Tricuspid Valve Re-replacement	3	0

Note. ASD – atrial septal defect; CCTGA – congenitally corrected transposition of the great arteries; LV – left ventricle; LVOT – left ventricular outflow tract; PA – pulmonary atresia; TGA – transposition of the great arteries; VSD – ventricular septal defect.

Table 21. RACHS-II Category 4 Risk Stratification for Congenital Heart Surgery (Cohort Aged 1–18 Years)

Procedure	Procedural Volume	Mortality, %
Correction of Truncus Arteriosus	9	0
Correction of TAPVR	2	0
Correction of Pulmonary Vein Stenosis	2	0
Correction of Anomalies of Venous Return	3	0
Take-down of Fontan Circulation	1	0
Resection of Coarctation and Pulmonary Artery Banding	2	0
Stenting of PDA	20	5
Creation of Systemic-to-Pulmonary Artery Shunt	16	0
Creation of RV-PA Shunt	4	0
Pulmonary Artery Banding (PAB)	12	0
Removal of Pulmonary Artery Band	20	0

Note. PDA – patent ductus arteriosus; RV-PA – right ventricle to pulmonary artery; TAPVR – total anomalous pulmonary venous return.

a significant proportion of these defects are operated on during the neonatal period or early infancy suggests that an effective system for patient transfer from maternity hospitals to cardiac surgery centers has been established in the Russian Federation. Nevertheless, the persisting mortality in the group of complex CHD necessitates further analysis of logistical chains: transport time, availability of prostaglandin therapy during evacuation stages, and the level of preoperative stabilization in regional centers.

The most sensitive indicator of the quality of a cardiac surgery service is mortality in the neonatal group (0–28 days) [13]. In our study, this rate was 6.6 % (102 patients out of 1,542), which is expectedly higher than in older age groups (2.1 % in infants under one year and 0.3 % in children older than one year). The more than twenty-fold disparity in mortality rates between neonates and children over one year (6.6 % vs. 0.3 %) underscores that neonatal cardiac surgery remains the area of greatest risk. This is attributable not only to the anatomical complexity of the defects (critical obstructions, single ventricles) but also to the physiological immaturity of the patients, low birth weight, and concomitant pathology [14]. The subgroup of low birth weight infants (<2.5 kg)

Table 22. RACHS-II Category 5 Risk Stratification for Congenital Heart Surgery (Cohort Aged 1–18 Years)

Procedure	Procedural Volume	Mortality, %
Damus – Kaye – Stansel Operation	3	0

deserves special attention, where mortality was 6.5 %. Although this rate does not exceed the average mortality for the neonatal group, surgical treatment in this cohort is associated with technical difficulties in cannulation, perfusion, and postoperative management. Analysis of cardiopulmonary bypass utilization in neonates revealed a mortality rate of 9.2 %. This constitutes the “core” of the problem: complex reconstructions (arterial switch operation, Norwood procedure, aortic arch reconstructions) performed under CPB contribute the majority of mortality. Comparison with STS Database data shows that in leading North American centers, mortality for STAT Category 5 procedures (highest complexity) ranges from 10 to 20 % [15; 16]. Our data for RACHS-II Category 5 (24.61 % overall mortality and 29.78 % for the Norwood procedure) demonstrate that, despite advances, the treatment of hypoplastic left heart syndrome (HLHS) remains a significant challenge for Russian medicine. The persisting mortality rates associated with the Norwood procedure necessitate a comprehensive analysis of risk factors. Key determinants of outcome are likely to include the influence of preoperative patient status, the surgical learning curve, and the capabilities for postoperative care across different institutions [17–20].

The application of the RACHS-II scale allowed for the objectification of the complexity of the interventions performed. Category 1 (low risk) demonstrated a mortality rate of 0.33 %. This is an excellent result, confirming the routine nature of procedures such as ASD closure or patent ductus arteriosus (PDA) ligation. However, even here, isolated fatalities occurred (e.g., for ASD closure – 0.06 %). This serves as a reminder that in cardiac surgery, no operation is entirely simple, and complications can arise even during standard procedures. Categories 2 and 3 (moderate risk) show acceptable figures (0.98 % and 6.47 %, respectively). The majority of radical repairs are represented in this group, most notably the correction of Tetralogy of Fallot, which carries a mortality rate of 0.6–4.4 % depending on the surgical approach. It is important to note the difference in outcomes between transannular patch repair (4.40 %) and preservation of the pulmonary valve annulus (0.60 %). This confirms the contemporary trend towards valve-sparing techniques, although the choice of method is often dictated by

anatomy rather than surgeon preference [21]. Categories 4 and 5 (high risk) are predictably associated with high mortality (7.17 % and 24.61 %). The data obtained for the arterial switch operation (7.9 % in Category 2 and 9.37 % in Category 3 with VSD) are somewhat higher than in leading international centers of excellence, where this rate approaches 2–3 %. However, the heterogeneity of the sample must be considered: the data combine both federal centers with a throughput of hundreds of operations and regional clinics with isolated cases of TGA. The well-documented phenomenon of the learning curve is clearly evident in pediatric cardiac surgery: centers with a high volume of complex interventions demonstrate superior outcomes.

One of the most striking results of the study is the proportion of endovascular interventions, reaching 38.4 % (5,619 procedures) with a minimal mortality rate of 0.3 %. This signifies a radical paradigm shift in the treatment of CHD in Russia. Closure of ASD and PDA, and balloon valvuloplasties have become the methods of choice, replacing open surgery where feasible [22; 23]. This not only reduces patient trauma and risks but also yields economic benefits through reduced hospital stays. The results of surgical treatment for cardiac arrhythmias warrant separate attention. In a cohort of 1,342 patients, no fatalities were registered (0 %), indicating a high safety profile for modern interventional procedures. Advances in clinical electrophysiology allow these techniques to be considered a means of radical correction for tachyarrhythmias, providing complete recovery and the possibility of discontinuing lifelong antiarrhythmic therapy [24].

Analysis of the volume of care by center (see Table 1) reveals a significant disparity. The top 5 centers (A.N. Bakulev National Medical Research Center for Cardiovascular Surgery (NMRCCS), E.N. Meshalkin National Medical Research Center (NMRC), Federal Centers for Cardiovascular Surgery in Astrakhan and Penza, V.A. Almazov NMRC) account for more than 40 % of all operations. The leading institution (A.N. Bakulev NMRCCS) performs 2,709 operations, while several regional hospitals perform fewer than 50–100 annually. The global debate on “centralization versus accessibility close to home” is also relevant for Russia [25]. On one hand, the concentration of complex cases (RACHS-II Categories 4–5) in federal centers ensures the accumulation of experience and improved outcomes. On the other hand, regional centers are vital for providing emergency care, performing stabilization phases, and conducting standard operations (Categories 1–2). The registry data indicate that the system operates on a mixed model. It is critically important that “smaller” centers adequately assess their

capabilities and promptly refer patients with complex defects to expert-level institutions.

Multi-stage hemodynamic correction for complex congenital heart diseases remains one of the most challenging areas of pediatric cardiac surgery [26; 27]. Analysis of the outcomes of the final stages of treatment presents an encouraging picture: hospital mortality for the Fontan operation (50 cases, 4.00 %) and the Glenn operation (104 cases, 3.84 %) is at a socially acceptable level, comparable to data from leading global clinics. These figures confirm that upon successful navigation of the neonatal period, the prognosis for patients with a single ventricle becomes favorable.

However, the problematic area and the primary source of patient loss remains the initial palliative stage [28]. The mortality rates for systemic-to-pulmonary artery shunt (SPS) creation, amounting to 9.74 % (Risk Category 4), are a concerning signal. Contrary to the common perception of the technical simplicity of shunt placement, this intervention requires meticulous intraoperative management of the balance between pulmonary and systemic blood flow (Qp/Qs). The high mortality in this group likely reflects the difficulties of postoperative stabilization in children with unstable hemodynamics.

In this context, a comparative analysis of surgical and interventional methods for securing pulmonary blood flow is of fundamental importance. Our data show that stenting of the patent ductus arteriosus in ductus-dependent patients (101 procedures) was associated with a mortality rate of 4.95 %, almost half that of surgical SPS creation (9.74 %). The effectiveness of the endovascular approach, especially in low birth weight and somatically compromised neonates, confirms the relevance of the global trend towards the adoption of hybrid strategies [29]. In this setting, PDA stenting serves not merely as an alternative but as a safer “bridge” to radical or subsequent palliative correction, allowing for minimization of surgical trauma during the critical period [30].

Study Limitations

Several limitations must be considered when interpreting the data. First, the study is retrospective in nature. Second, while the use of the RACHS-II scale represents a step forward, it assesses only procedural risk, without fully accounting for patient comorbidity (prematurity, genetic syndromes, extracardiac pathology). A patient with a simple ventricular septal defect but severe pneumonia and renal failure has a significantly higher risk than a healthy child with the same defect, yet RACHS-II equates them. Furthermore, some incompleteness was observed in the database regarding the “primary/repeat” operation classification (the sum of percentages does

not equal 100%), indicating a need for improved medical record-keeping and local data entry into the registry. Another important aspect is the definition of mortality. We utilized 30-day and in-hospital mortality. However, in contemporary cardiac surgery, 90-day mortality and long-term survival are gaining increasing significance, as, for example, following the Norwood procedure, a significant proportion of fatalities occur during the interstage period, after hospital discharge.

Conclusion

The conducted analysis of data from the registry of the Interregional Public Organization of Pediatric Cardiac Surgeons for the year 2024 demonstrates that

surgical care for children with congenital heart diseases in the Russian Federation is provided at a high level. The overall postoperative mortality rate does not exceed 1.42%, which aligns with contemporary international standards. Key factors influencing the outcomes of surgical treatment include patient age, the type and complexity of the surgical intervention, and the utilization of cardiopulmonary bypass.

The obtained results also indicate the necessity for further development of perinatal diagnostics, improvement of surgical technologies, particularly in regional medical centers with CHD departments, to ensure the accessibility of high-technology care.

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