



# Complications After Surgical Treatment Of Anomalous Pulmonary Venous Connection

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**ABSTRACT**

Anomalous pulmonary venous connection (APVC) is a congenital heart defect in which one or more pulmonary veins drain into the right atrium or systemic veins instead of the left atrium. This results in oxygenated blood returning to the right heart chambers, where it mixes with deoxygenated venous blood. Without timely surgical intervention, APVC leads to chronic hypoxia and progressive heart failure, affecting the child's development and survival. Despite successful surgical correction, certain complications may arise in the postoperative period, requiring prompt attention. Literature analysis indicates that studying the outcomes and complications of APVC correction remains a highly relevant clinical issue. The aim of this study was to conduct a comparative evaluation of surgical outcomes for APVC correction depending on the type of defect and to identify the causes of mortality in the early postoperative period.

**Keywords:**

congenital heart defect, anomalous pulmonary venous connection, complications, outcome assessment

**Relevance.** Anomalous pulmonary venous connection (APVC) is a congenital heart defect in which one or more pulmonary veins connect to the right atrium or to the systemic venous circulation instead of draining normally into the left atrium [1,4,6,10,14,23]. This results in oxygenated blood from the lungs returning to the right side of the heart, where it mixes with deoxygenated venous blood. In 90% of cases, anomalous pulmonary venous connection is associated with an atrial septal defect (ASD). Depending on the number of anomalously draining pulmonary veins, partial anomalous pulmonary venous connection (PAPVC) is distinguished—when one or several, but not all, pulmonary veins drain into the right heart—and total anomalous pulmonary venous connection (TAPVC), when all pulmonary veins drain into the right heart chambers. Without timely surgical correction, anomalous pulmonary venous connection leads to chronic hypoxia and progressive heart failure, affecting

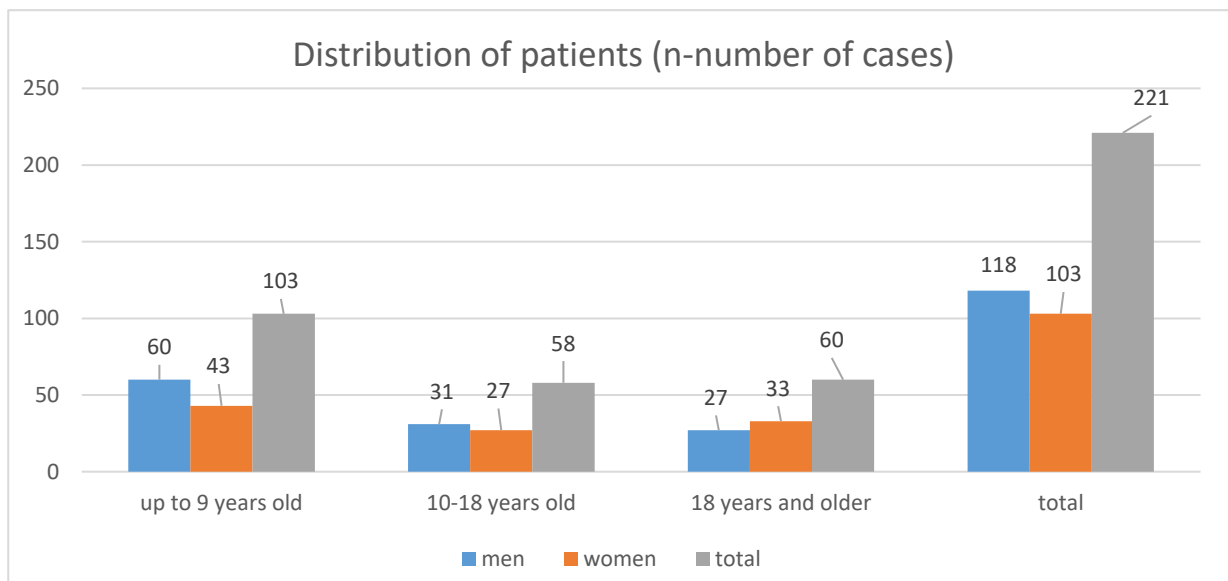
the child's development and survival. Despite successful surgical intervention, certain complications may develop in the early postoperative period and require immediate attention. A review of the literature indicates that the study of outcomes and complications after surgical correction of this defect remains highly relevant [2,5,8,9,12,15,20]. Therefore, further investigation of this issue has important practical and scientific significance. Objective of the study. To conduct a comparative assessment of the outcomes of surgical correction of anomalous pulmonary venous connection depending on the type of defect, and to identify the causes of mortality in the early postoperative period.

**Materials and Methods**

From 2001 to 2021, a total of 221 patients underwent surgical correction of anomalous pulmonary venous connection at the Republican Specialized Scientific and Practical

Medical Center of Surgery named after Academician V. Vakhidov and at the clinic of Andijan State Medical Institute. The patients' age ranged from 0.1 to 45 years, with a mean

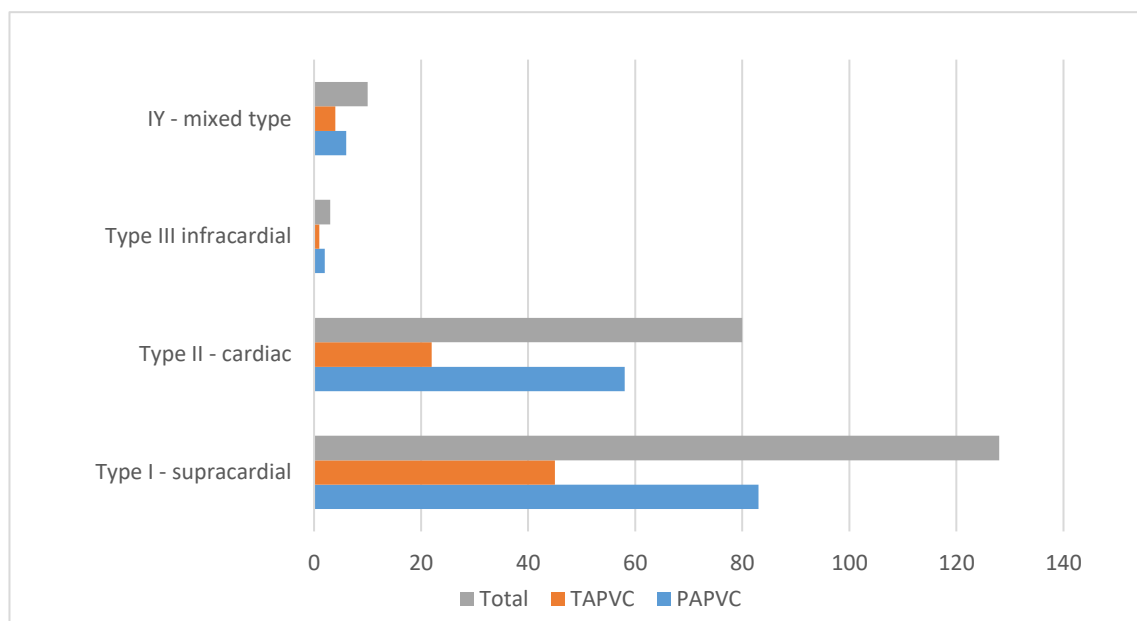
age of  $16.3 \pm 0.26$  years. Among the operated patients, 118 (53.3%) were male and 103 (46.7%) were female (Fig. 1).



**Fig.1. Distribution of patients by sex and age**

Based on clinical and instrumental examination data, total anomalous pulmonary venous connection (TAPVC) was identified in 72 patients (32.5%), while partial anomalous pulmonary venous connection (PAPVC) was diagnosed in 149 patients (67.5%). All patients were divided into groups according to the generally accepted classification of Richard Darling et al. [18] (Fig. 2).

All patients underwent a comprehensive clinical evaluation, including assessment of complaints and medical history, physical examination, electrocardiography, phonocardiography, chest radiography, and laboratory investigations.



**Fig.2. Distribution of patients according to the type and anatomical variant of the defect**

In addition, special diagnostic methods were used, including echocardiography, multislice computed tomography, cardiac catheterization with angiography, and intraoperative tensiometry.

All types of defect correction were performed under cardiopulmonary bypass. In most cases (86.5%), open-heart surgery was carried out via median sternotomy, while in 20 patients (13.4%) a right lateral thoracotomy approach was used. Cannulation of the aorta and venae cavae was performed according to the standard protocol.

Postoperative complications were conditionally divided into early and late according to the time of onset. Early postoperative complications, occurring within the first hours and days after surgery, were observed in 42 patients (19.0%). The nature and frequency of these complications are presented in Figures 3 and 4.

According to the literature, the main potential complications can be grouped as follows:

1. Recurrent pulmonary vein stenosis. Renarrowing at the site of the created anastomosis or within the venous collector itself. This is the most frequent and serious long-term complication, especially in infants. Clinical signs include progressive dyspnea, tachycardia, decreased oxygen saturation, and poor weight gain. Early diagnosis using echocardiography and, when necessary, computed tomography angiography is critically important.

2. Cardiac arrhythmias. Rhythm disturbances, such as supraventricular tachyarrhythmias, may occur due to irritation of the cardiac conduction system during surgery. These are usually managed medically.

3. Infectious complications. Pneumonia, postoperative wound infection, or sepsis may develop despite preventive measures and require prompt antibacterial therapy.

4. Pleural and pericardial effusion. Accumulation of fluid in the pleural cavity or pericardial space may lead to respiratory insufficiency or impaired cardiac function. Drainage may be required.

5. Chylothorax. A rare but serious complication characterized by accumulation of lymphatic

fluid in the pleural cavity due to injury of lymphatic vessels during surgery. It requires specialized treatment and dietary management.

6. Persistent pulmonary hypertension. In cases with severe, irreversible changes in the pulmonary vasculature prior to surgery, elevated pulmonary artery pressure may persist after correction of anomalous pulmonary venous connection, requiring additional targeted therapy.

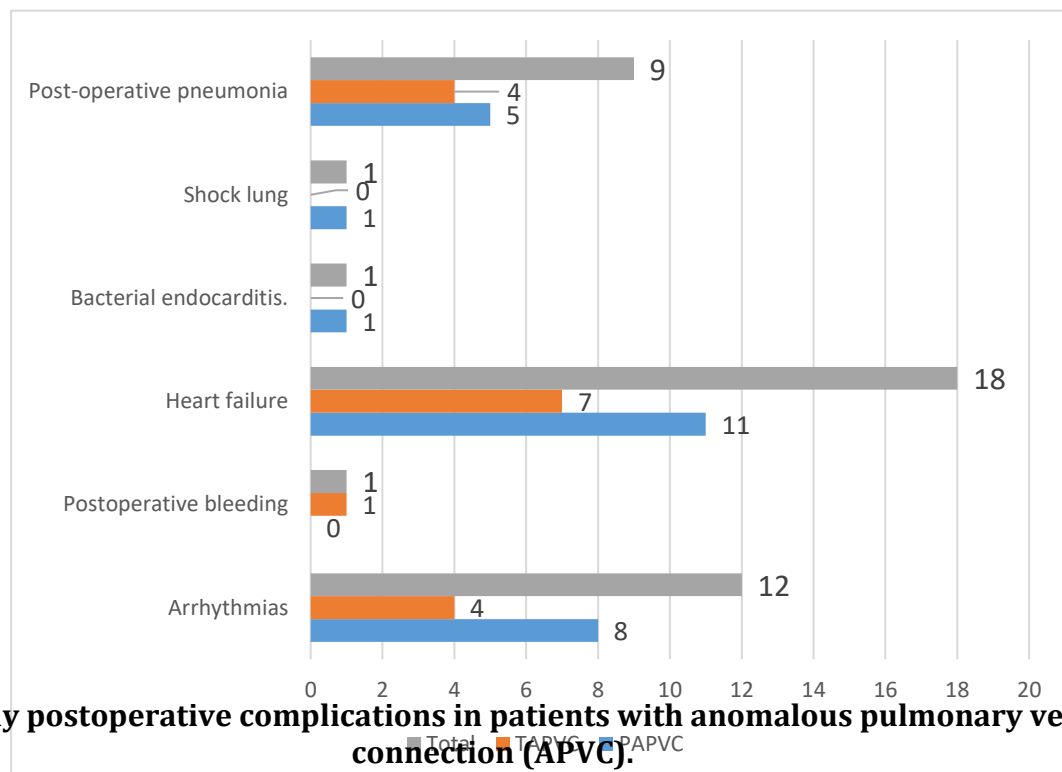
Medical staff must carefully monitor for signs of these complications to ensure timely treatment. Complications related to technical errors during surgical correction. This group includes inadequate correction of the defect, intraoperative bleeding, and injury to the cardiac conduction system during the intracardiac stage of surgery (Fig. 3). Among the operated patients with total anomalous pulmonary venous connection (TAPVC), intraoperative bleeding occurred in 1 patient (0.4%) and was promptly controlled. In 12 patients (5.4%) with anomalous pulmonary venous connection (APVC), various types of arrhythmias were observed, including bigeminy, ventricular extrasystoles, ventricular rhythm, and atrioventricular block. All of the above-mentioned arrhythmias resolved after appropriate medical therapy.

To date, the literature continues to report intraoperative errors (incomplete correction of the defect) and late postoperative complications, such as septic endocarditis leading to partial or complete recanalization of the defect. The possibility of defect recanalization has been reported by many authors [7,10,11,16,19,23]. Another group of specific late complications is associated with impaired blood flow through the anomalously draining pulmonary veins due to stenosis or thrombosis [11,13]. According to F. Lewis et al. [21], signs of thrombosis of anomalous pulmonary venous connection with features of lobar pulmonary congestion—predominantly of the right upper lobe—and hemoptysis in one patient were observed in four cases. Such a complication rarely leads to death but does not resolve without consequences, as it may result in the formation of adhesions and collateral circulation. Blood from the affected lobe may

then drain into the systemic veins, leading to recurrence of the left-to-right shunt [3,14,18,24].

Complications related to the natural course of the defect (Fig. 3) include heart failure and bacterial endocarditis, which were observed in 19 patients (8.5%)—1 case of bacterial

endocarditis and 18 cases of heart failure of varying severity. In all cases, appropriate therapeutic measures resulted in a favorable outcome. Bacterial endocarditis that developed in the early postoperative period was successfully treated conservatively.

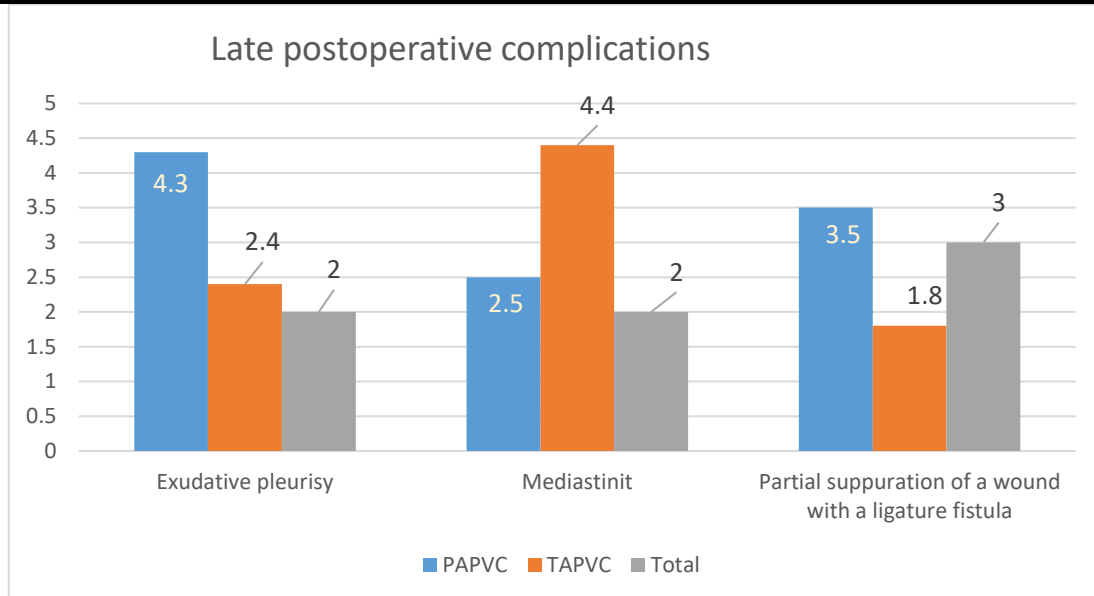


**Fig. 3. Early postoperative complications in patients with anomalous pulmonary venous connection (APVC).**

Complications associated with cardiopulmonary bypass and mechanical ventilation were observed in two patients: one patient (0.4%) developed acute respiratory distress syndrome (“shock lung”), and another developed obstructive pneumonia with respiratory failure (Fig. 3). No concomitant diseases complicating the postoperative course were observed in our series.

Late postoperative complications during the hospital period were observed in 19 patients (8.6%) (Fig. 4): exudative pleuritis in 3 patients (1.36%); late wound suppuration with the development of mediastinitis in 4 patients (1.8%); and partial wound infection with formation of a ligature fistula in 1 patient

(0.4%). All these patients received appropriate treatment and procedures in the department and were subsequently discharged home in satisfactory condition. There are also specific complications associated with impaired blood flow through the superior vena cava (SVC) due to various causes, including narrowing of its lumen after division, shrinkage of patches, or thrombosis. S. Schuster et al. [22] reported signs of temporary SVC obstruction in one patient. C. Chartrand et al. [16] described SVC stenosis detected five years after correction of anomalous pulmonary venous connection in a patient with a persistent left SVC. V.S.Sergievskiy et al. [11] reported a case of thrombosis leading to cerebral edema.



**Fig. 4. Late postoperative complications in patients with anomalous pulmonary venous connection (APVC).**

Impaired blood flow through the superior vena cava (SVC) may have a favorable outcome and be compatible with life [3,17], despite narrowing or even complete obstruction [21]. In such cases, venous return is maintained through collateral circulation via the azygos vein system into the inferior vena cava, or through a persistent left SVC when present.

Despite successful surgical intervention, some patients may develop late complications requiring further medical attention. Awareness of these risks allows for timely detection and prevention of progression. A review of the literature indicates that the most significant late complications include:

1. Restenosis (recurrent narrowing) of the pulmonary veins. This is the most common and serious late complication, especially in infants who have undergone correction of obstructive total anomalous pulmonary venous connection (TAPVC). Restenosis may occur due to scar formation at the anastomotic site or intimal proliferation. Clinically, it presents with recurrent dyspnea, tachycardia, decreased oxygen saturation, and poor weight gain
2. Persistent or recurrent pulmonary hypertension. In cases where severe and irreversible pulmonary vascular changes

developed before surgery, pulmonary hypertension may persist after correction of anomalous pulmonary venous connection (APVC). It may also recur in the presence of pulmonary vein restenosis.

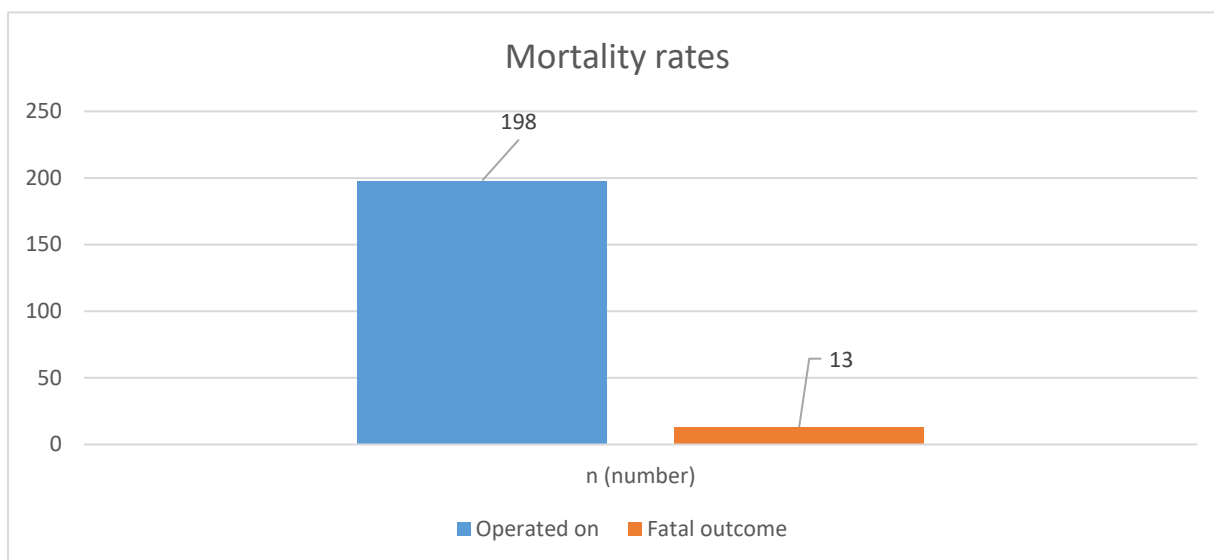
3. Cardiac arrhythmias. Some patients may experience rhythm disturbances, particularly atrial arrhythmias associated with postoperative atrial scarring.

4. Right ventricular dysfunction. If the right ventricle was subjected to prolonged and significant volume overload before surgery, its function may not fully recover.

5. Residual atrial septal defect. In rare cases, a small residual interatrial shunt may remain after ASD closure. It is usually not clinically significant but requires follow-up.

6. Residual symptoms. Some children may experience mild dyspnea or fatigue during intense physical exertion even after successful surgery.

Hospital mortality in our series was 5.9% (13/221). It should be noted that all fatal outcomes occurred after correction of total anomalous pulmonary venous connection (TAPVC) (Fig. 5). In recent years, this rate has been reduced to a minimum in the latest operations for APVC.



**Fig. 5. Mortality rates**

Analysis of the causes of unfavorable outcomes revealed that mortality was not always attributable to technical surgical errors. In our observations, surgical outcomes in patients with TAPVC largely depended on age and the initial severity of the condition. Mortality was observed primarily in patients operated on during early infancy and in children with severe pulmonary hypertension.

Characterizing the causes of mortality overall is challenging, as they depend on the anatomical type of the defect, the degree of pulmonary hypertension, and the age of the patients. Surgical technical skill also plays a significant role. In our series, the causes of fatal outcomes

were as follows: In 3 cases, acute heart failure was the primary cause of death. In 2 cases, acute heart, respiratory, and renal failure developed, leading to grade III cerebral coma due to homologous blood syndrome. In 1 case, death resulted from cerebral air embolism, cerebral edema, grade III coma, and cardiovascular and renal failure, considered a complication of cardiopulmonary bypass. In 4 cases, acute cardiovascular failure occurred due to arrhythmogenic shock, which led to death. In 3 cases, acute heart failure developed postoperatively, was unresponsive to treatment, and caused death (Table 5).

**Causes and timing of death of patients with TADPV (n=13). Table 5**

Diagnosis	Number of deaths (n-13)	Cause of death	Dates of death
Type II TAPVC	2	Acute cardiorespiratory and renal insufficiency. Grade III cerebral coma.	Day 4
Type II TAPVC	1	Acute cerebrovascular accident. Brain embolism. Swelling of the brain. Cerebral coma of the third art. Heart vascular failure. Kidney failure	Day 4
Type II TAPVC	3	Acute cardiac vascular insufficiency. Swelling of the brain.	Day 3
Type I TAPVC	4	Acute cardiovascular insufficiency. Arrhythmogenic shock.	Day 1
Type I TAPVC	3	Acute cardiovascular insufficiency	Day 9

After successful surgical correction of anomalous pulmonary venous connection, patients enter a phase of long-term recovery and outpatient follow-up, aimed at ensuring the child's full and healthy development. Despite the radical elimination of the defect, former patients require regular medical monitoring, as there remains a risk of late complications. The long-term prognosis after surgery is generally favorable, but it demands careful adherence to medical recommendations and prompt response to any changes in health status.

The overall prognosis for children who have undergone surgical correction of anomalous pulmonary venous connection has significantly improved due to advances in cardiac surgery and intensive care. In most cases, successful intervention restores normal pulmonary venous return and eliminates the volume overload of the right heart chambers, leading to normalization of cardiac function and resolution of systemic hypoxia symptoms. Many patients who have undergone APVC correction are able to live full, active lives, achieving normal growth and development without significant limitations in physical activity. However, it is important to recognize that lifelong cardiological follow-up is necessary.

Based on the above, it is evident that the treatment of anomalous pulmonary venous connection, particularly in its total form, still faces several unresolved challenges. These include the risk of superior vena cava stenosis, impaired pulmonary venous outflow, persistence of left-to-right shunting, and cardiac rhythm disturbances.

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