



## OPEN Prognostic factors in pediatric TAPVC: a 10-year retrospective study

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Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart defect where the pulmonary veins abnormally connect to the systemic venous circulation, rather than the left atrium (LA). This study retrospectively analyzed 48 children diagnosed with TAPVC from January 2014 to January 2024 to investigate how specific echocardiographic and morphological characteristics impact prognosis. Perioperative records, including echocardiography, computed tomography angiography (CTA) images, surgical details, and patient outcomes, were reviewed. Data were statistically analyzed using tools such as t-tests, Mann-Whitney U tests,  $\chi^2$  tests, and logistic regression. Key factors examined included age at surgery, anatomical subtype of TAPVC, presence of preoperative obstruction, surgical techniques used, and postoperative complications. By identifying the impact of these variables on survival rates and hospital recovery, the study aims to improve risk stratification and management strategies for better outcomes in children with TAPVC.

**Keywords** Total anomalous pulmonary venous connection, Echocardiography, Pulmonary venous obstruction, Prognosis

### Abbreviations

TAPVC	Total anomalous pulmonary venous connection
LA	Left atrium
RA	Right atrium
PVO	Pulmonary venous obstruction
CPV	Common pulmonary vein
CS	Coronary sinus
SVC	Superior vena cava
ASD	Atrial septal defect
PFO	Patent foramen ovale
VSD	Ventricular septal defect
PDA	Patent ductus arteriosus
PS	Pulmonary stenosis
PASP	Pulmonary arterial systolic pressure
CTA	Computed tomography angiography

Total anomalous pulmonary venous connection (TAPVC) is a relatively rare and severe cyanotic congenital heart disease, occurring in approximately 0.5–2% of congenital heart diseases<sup>1</sup>. In this condition, all pulmonary veins fail to connect to the left atrium (LA); instead, they connect to the right atrium (RA) through systemic veins or drain directly into the RA. Based on the drainage pathways, TAPVC can be divided into four types: supracardiac, cardiac, infracardiac, and mixed. The supracardiac type is the most common, representing about 43% of all cases. Additionally, about 25–50% of patients with TAPVC have associated pulmonary venous obstruction (PVO)<sup>2–4</sup>.

The clinical symptoms of children with TAPVC are closely related to the presence of PVO, these symptoms can range from mild pulmonary hypertension to severe pulmonary congestion and acidosis<sup>5</sup>. In case where PVO is present, emergency surgery is required. Without timely intervention, most affected neonates do not survive the neonatal period.

Advancements in surgical techniques, diagnostic procedures, and perioperative care have significantly reduced mortality rates in TAPVC in recent years. Various studies have focused on prognostic factors such

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as preoperative general conditions (age, weight, oxygen saturation, pH value), surgical techniques, and cardiopulmonary bypass time<sup>6</sup>. While much of the research has concerned on the type and presence of PVO, less attention has been given to specific echocardiographic parameters.

This study aims to bridge that gap by retrospectively analyzing the correlation between detailed echocardiographic measurements and the prognosis of children with TAPVC. By understanding these relationships, better predictive models and tailored management strategies can be developed, potentially improving outcomes for patients with TAPVC.

Results

Clinical characteristics

In this cohort of 48 children diagnosed with TAPVC, the median age was 35 days (range 1-2615days), and the median weight was 3.8 kg (range 1.5–13.5 kg). The median surgical duration for these cases was 215 min, with a range from 135 to 485 min. The gender distribution included 34 males (approximately 70.8%) and 14 females (approximately 29.2%). Among of TAPVC connections, 15 cases (31.3%) were supracardiac, 20 cases (41.7%) were cardiac, 10 cases (20.8%) were infracardiac, and 3 cases (6.3%) had mixed connections. These details of the patient demographics and clinical characteristics in the study were shown in Table 1.

The drainage pathway of TAPVC

All 48 cases underwent echocardiographic examination to evaluate the pulmonary venous drainage pathways, the presence of obstruction, associated malformations, and cardiac function. Among the 15 supracardiac types, nine cases drained into the innominate vein and six into the superior vena cava (SVC) (Fig. 1). There were 13 single vertical vein and 2 cases of double vertical veins. In the 20 cardiac cases, 17 CPVs drained into the RA via the coronary sinus (CS) (Fig. 2), two CPV drained directly into the RA, and one had four pulmonary veins draining separately into the RA. Among the 10 infracardiac cases, eight cases had CPVs that drained into the portal vein via a vertical vein, one drained into the hepatic vein, and one drained into the inferior vena cava via ductus venosus (Fig. 3). In the three cases mixed-type cases, all of them combines both supracardiac and cardiac types (Fig. 4).

Outcome of the surgery

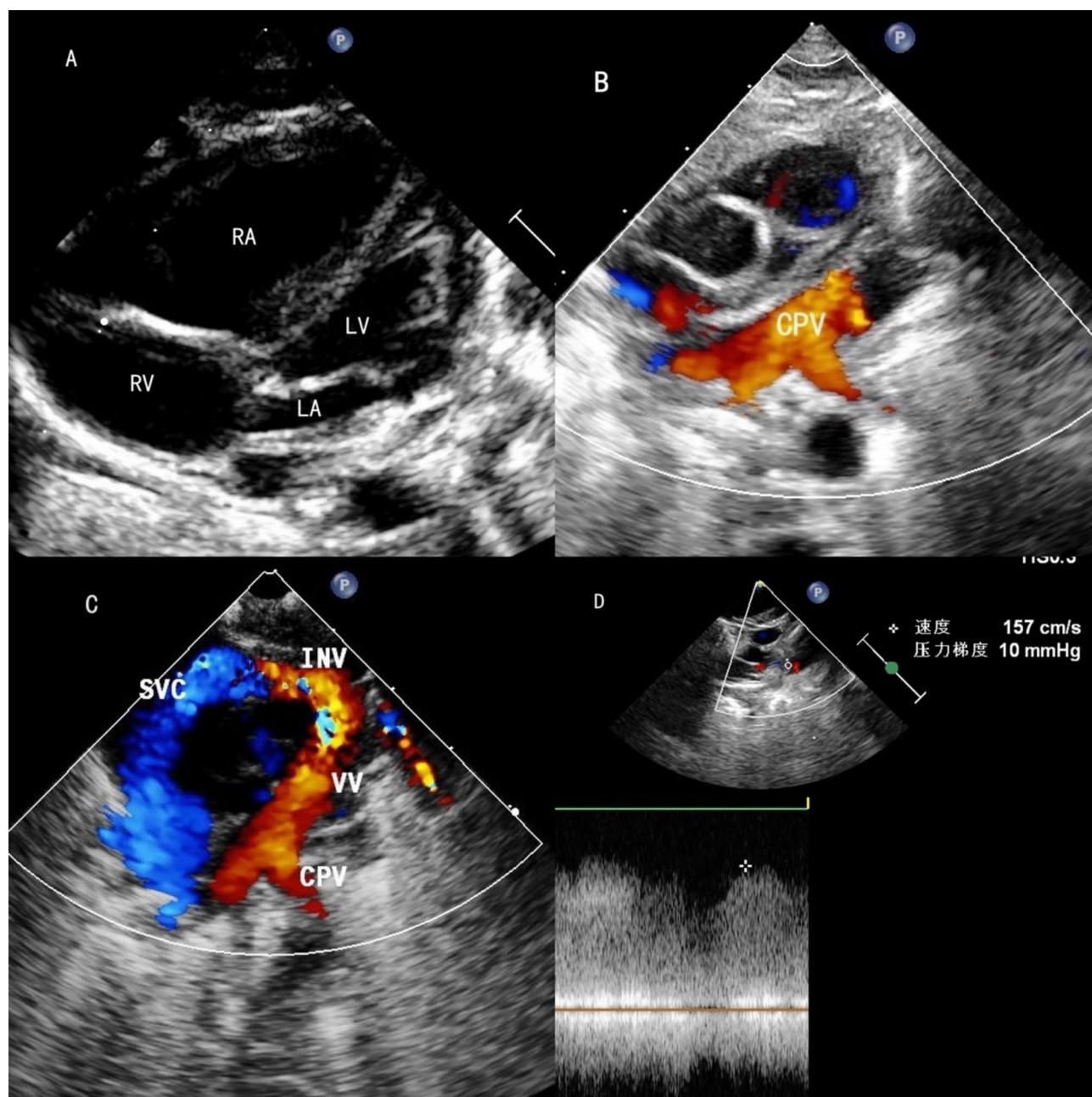
Postoperative death occurred in 14 (29%,14/48) cases, distributed as follows: supracardiac (26.7%,4/15), cardiac (20%,4/20), infracardiac (40%,4/10), mixed (66.7%,2/3). Postoperative PVO occurred in 14 cases (29%, 14/48), with the following distribution: supracardiac (20%,3/15), cardiac (10%,2/20), infracardiac (60%,6/10), mixed (100%,3/3). Among the 14 deaths, 11 patients had preoperative PVO, while three did not. Other postoperative complications included diaphragmatic paralysis in 2 cases, SVC obstruction in 1 case, and bronchial stenosis in 1 case.

The echocardiographic characteristics of TAPVC

The postoperative outcomes of patients with TAPVC were analyzed concerning various preoperative anatomical and physiological parameters, including drainage pathways, the diameter of the CPV, the size of the LA, the

Patient characteristics	
Gender (male/female)	34/14
Surgical age (d)	35 days (1-2615 days)
Weight (kg)	3.8 kg (1.5–13.5 kg)
Hospital stays (day)	32 days (2-117 days)
Operative time (min)	215 min (135–485 min)
Associated cardiac lesion	
PDA	23
ASD/PFO	48
VSD	4
Mild PS	3
Anatomic type	
Supracardiac	15
Cardiac	20
Infracardiac	10
Mixed	3
Preoperative PVO	15
Associated extracardiac abnormality	
Congenital horseshoe lung, hypoplasia of the right lung, scoliosis	1
Esophageal atresia and esophagotracheal fistula	1

**Table 1.** Patient characteristics. ASD atrial septal defect, VSD ventricular septal defect, PFO patent foramen ovale, PDA patent duct arteriosus, PS pulmonary stenosis, PVO Pulmonary venous obstruction.

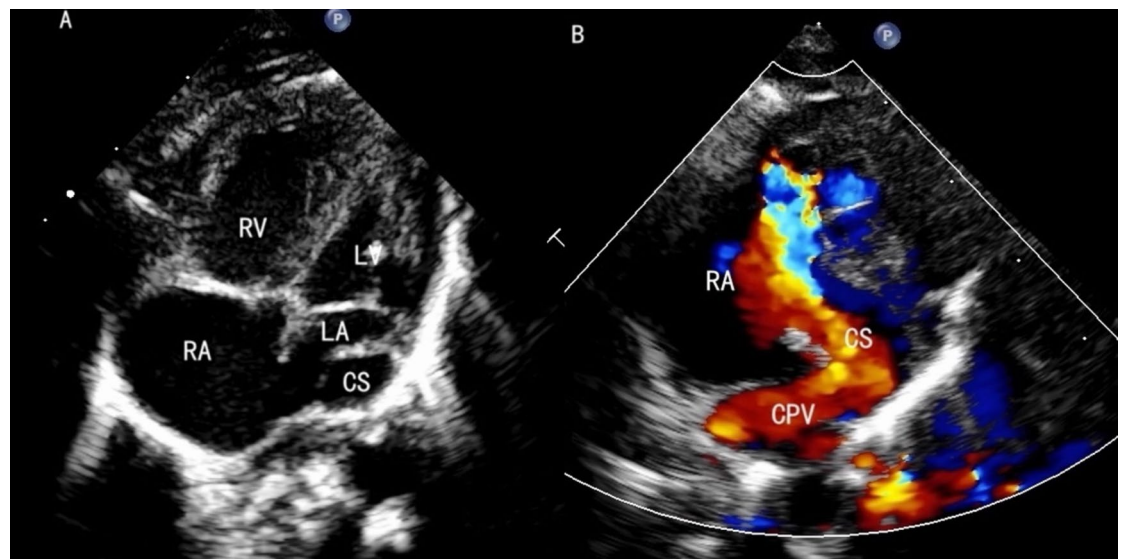


**Fig. 1.** The echocardiography of supracardiac TAPVC. (A) The left heart is extreme small. (B) The pulmonary veins drain into the CPV. (C) The CPV drains into the innominate vein via vertical vein. (D) An obstruction was detected at the drainage site.

patent foramen ovale (PFO) or atrial septal defect (ASD), the presence of a patent ductus arteriosus (PDA), the degree of pulmonary hypertension, and the associated intracardiac malformations. The significant findings revealed that a smaller size of the LA and preoperative PVO were strongly associated with higher risks of postoperative PVO and death ( $P < 0.05$ ) (Tables 2 and 3). When the LA is less than or equal to 0.85 cm, the risk of poor prognosis increases. Conversely, the drainage pathway, pulmonary arterial systolic pressure (PASP), CPV diameter, PFO, and the presence of PDA did not show significant associations with postoperative outcomes.

## Discussion

Echocardiography is the first line diagnostic modality for congenital heart diseases, including TAPVC. However, there are some shortcomings of echocardiography. Due to the narrow diameter or tortuous pathways of the pulmonary veins or vertical veins, echocardiography may not visualize them. In contrast, computed tomography angiography (CTA) has advantages over echocardiography in visualizing the obstructed veins and displaying atypical vessels<sup>7</sup>. In our study, there were two cases misdiagnosed by echocardiography: one case of supracardiac



**Fig. 2.** The echocardiography of cardiac TAPVC. (A) The CS is dilated behind the small LA. (B) The CPV drains into the CS.

was misdiagnosed as the mixed type (supracardiac and intracardiac), and another case of supracardiac TAPVC was misdiagnosed as partial supracardiac. Both were correctly diagnosed by CTA. Echocardiography offers distinct advantages, such as assessing blood flow velocity in pulmonary and vertical veins, evaluating cardiac function, and measuring pulmonary artery pressure without radiation exposure- capabilities CTA lacks. Additionally, echocardiography excels at observing the size of the PFO or ASD and detecting the presence of a PDA. However, CTA is superior in accurately evaluating the course and number of anomalous veins, making it particularly effective for visualizing atypical vessel structures<sup>8,9</sup>. Therefore, combining both echocardiography and CTA provides a comprehensive preoperative evaluation by leveraging the strengths of each modality.

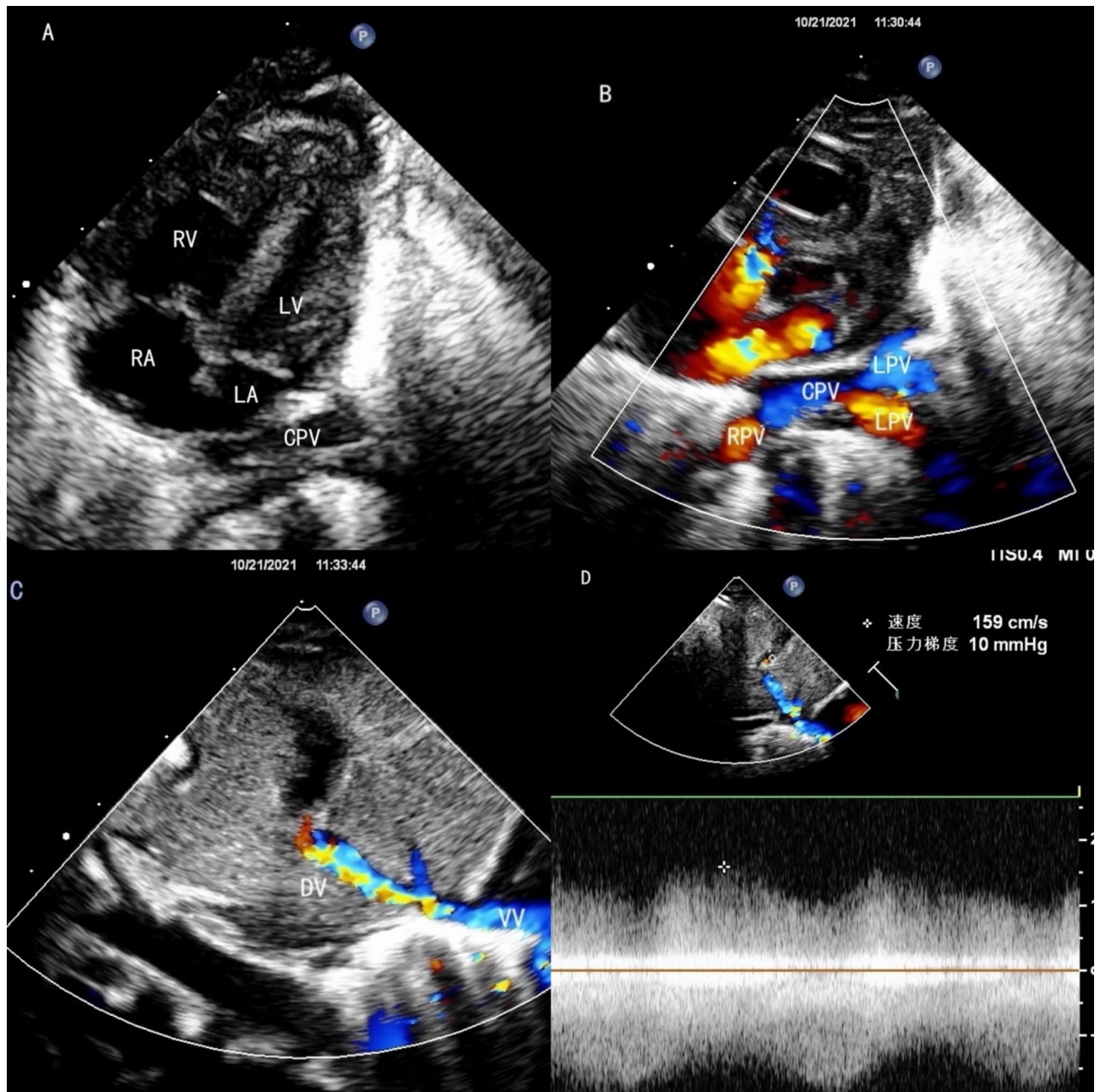
The drainage pathway of pulmonary veins in TAPVC is a debated prognosis factor. Most studies indicate that infracardiac and mixed-type pathways pose higher risks for death and postoperative PVO, while supracardiac and cardiac types generally have better outcomes<sup>6,10–12</sup>. However, a retrospective in Toronto found that the cardiac type is a risk factor for postoperative death<sup>3</sup>. Additionally, patients with pulmonary veins draining into the coronary sinus (CS) are susceptible to postoperative PVO due to the accumulation of the neointimal tissue during intracardiac repair<sup>13</sup>. Specific morphological features are associated with postoperative PVO in patients with cardiac TAPVC, the confluence to total pulmonary vein ratio, length of drainage route, and pulmonary veins return to the RA roof are predictor for postoperative PVO<sup>14</sup>. This complexity underscores the need for tailored surgical and postoperative strategies to enhance patient prognosis. Mixed-type TAPVC features a wide anatomic variation including bilateral symmetrical asymmetrical connections, bilateral asymmetrical connections, and bizarre anatomic variants, with the later significantly increasing the risk of death, reportedly by 5.85 times<sup>15</sup>. Supracardiac TAPVC, the most common subtype, typically involves drainage into the innominate vein via left vertical vein (type-Ia), or into SVC or the right azygos vein (type-Ib). The type Ib connection is recognized as a risk factor for postoperative PVO and death<sup>16</sup>. Given this morphologic heterogeneity, individualized treatment and surgical planning are essential to mitigate risks and optimize patient outcomes.

Complexed cardiac lesion that needs staged intervention is an independent risk factor for mortality. However, when the associated cardiac lesion was repaired during TAPVC surgery, it was not a risk factor for death<sup>17</sup>. The intracardiac malformations in our study were VSD, PDA and mild PS, which can typically be managed effectively during the correction of anomalous pulmonary venous connection. Additionally, the heterotaxy syndrome has been recognized as a predictor of postoperative mortality in TAPVC<sup>18</sup>. Therefore, in this study, the associated cardiac malformation is not a significant prognosis factor of postoperative mortality.

Our findings reaffirm previous research that preoperative PVO significantly impacts survival rates in patients undergoing surgical correction of TAPVC<sup>3,6,19–21</sup>. Specifically, with PVO, only 6 out of 15 cases survived (40%), whereas without PVO, 26 out of 33 cases survived (approximately 78.8%), a statistically significant difference underscoring the increased risk associated with PVO. These results highlight the necessity for early detection and management of PVO, meticulous preoperative evaluation, and possibly modified surgical techniques to improve outcomes. This data is crucial for clinicians to refine protocols and enhance patient care strategies, aiming for higher survival rates and better overall outcomes in TAPVC surgeries.

PVO, especially when manifested during the fetal period, critically impacts the pulmonary vascular bed, leading to increased pulmonary vascular pressure, vascular remodeling, or even atresia of the pulmonary veins<sup>22,23</sup>. The intricate anatomy of PVO, which can be due to intrinsic factors within the veins or extrinsic factors like surrounding structures or a dilated heart, adds complexity to its management<sup>24</sup>. PVO may present as segmental stenosis or diffuse narrowing, at various locations and involving single or multiple sites, making accurate diagnosis challenging<sup>13,25,26</sup>. Advanced imaging techniques such as CTA are superior to echocardiography in



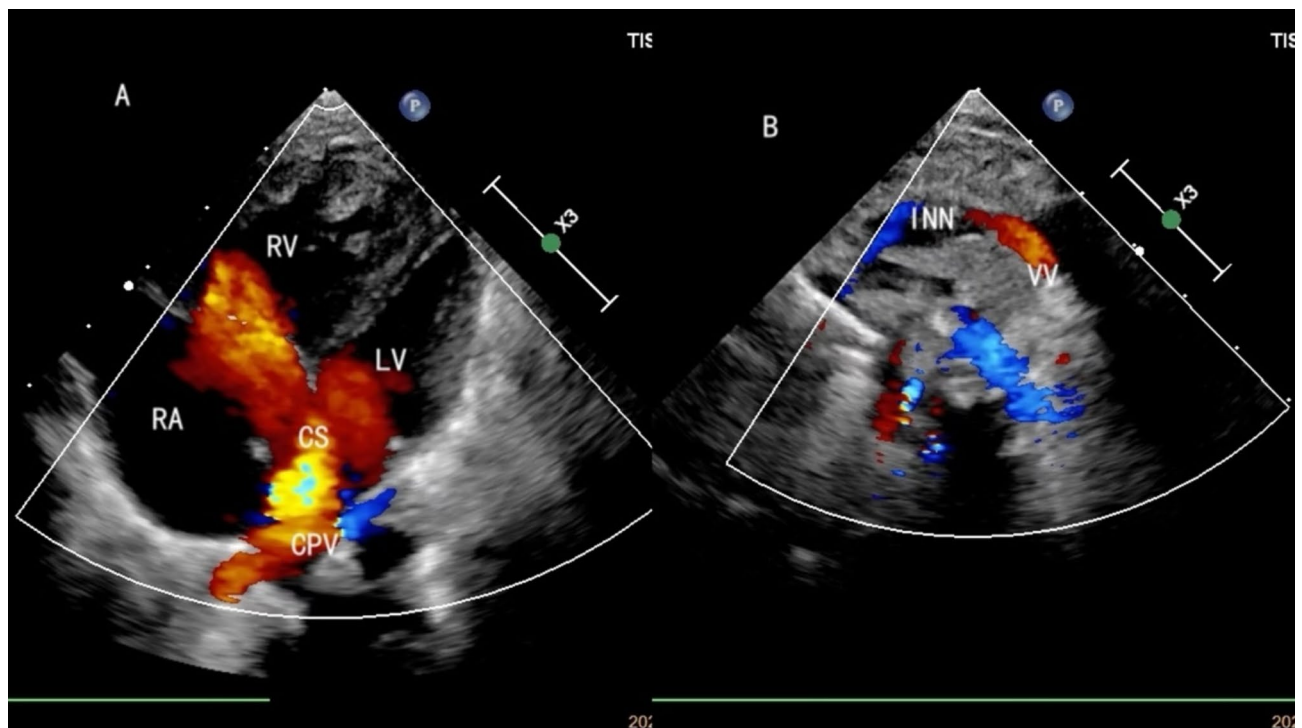


**Fig. 3.** The echocardiography of infracardiac TAPVC. **(A)** The left heart is small, the posterior wall of the LA is smooth, and the CPV is behind the LA. **(B)** The pulmonary veins drain into the CPV. **(C)** The vertical vein goes down then drains into duct vein, the color doppler shows turbulent flow at the drainage site. **(D)** A non-phasic flow was detected at the drainage site.

identifying these irregularities, underscoring the need for early and precise diagnostic interventions. To improve clinical outcomes, further research is necessary to optimize the assessment and management strategies for anomalous pulmonary veins in TAPVC patients.

In patients with TAPVC, the size of LA is crucial for assessing the development of the left heart, as blood flow to the left heart depends on right-to-left shunts, primarily including ASD, VSD, or PDA. While VSD and PDA often have bidirectional shunts, ASD typically involves a right-to-left shunt, with larger shunts facilitating better left heart filling and development. A small LA may lead to left heart overload postoperatively, poor pulmonary venous drainage, pulmonary congestion, and hypertension. Accurate preoperative evaluations of LA size and shunt sizes are necessary for surgical guidance and prognosis assessment. Our study highlighted that a small LA is an independent risk factor for death post-surgery, though the small sample size rendered ASD or PFO diameters non-significant as postoperative death risk factors.

Postoperative PVO is the major complication and the reason for reoperation, it also has impact on long term outcome. So it's urgent to prevent PVO after TAPVC repairment. The sutureless repair has been adopted



**Fig. 4.** The echocardiography of mixed TAPVC. (A) The CPV drains into RA via CS. (B) The other pulmonary vein drains into innominate vein via vertical vein.

to reduce postoperative PVO since 1990. But the contribution of sutureless technique in preventing subsequent PVO remain controversial. Its benefits over conventional technique needs further confirmation<sup>6,27</sup>.

## Methods

### Study population

This retrospective study analyzed data from 48 children diagnosed with TAPVC by echocardiography and confirmed by surgery at Fujian Maternal and Children's Hospital and Fujian Children's Hospital from January 2014 to January 2024. The study included children potentially with ASD, PFO, VSD, PDA, and mild PS, while excluding those with functionally univentricular circulation or atrial isomerism. This study was approved by the local ethics committee at Fujian Maternal and Children's Hospital.

### Ethics statement

The study was conducted in accordance with the Declaration of Helsinki and was approved by the Ethics Committee of Fujian Maternal and Child Health Hospital. The study subjects as well as their parents /legal guardians were informed about the study details and gave written consents to participate.

### Echocardiographic equipment and assessment

Echocardiographic imaging was performed by Philips IPEIQ 7C and Philips IE33 ultrasound diagnostic system with a 3–8 MHz phased-array probe. The sonographers assessed the drainage pathways of pulmonary veins, the presence of any obstruction, and the associated cardiac lesions.

A diagnosis of preoperative PVO was made based on echocardiography data indicating a non-phasic flow velocity greater than 1.5 m/s<sup>13</sup>. LA maximum diameter was measured at the end systole on the parasternal long-axis view. The ASD or PFO diameter was measured on the subcostal long-axis view. The CPV was measured on the maximum section. The PASP was evaluated by tricuspid regurgitation and divided into mild (30–50mmHg), moderate (51–70mmHg) and severe (> 70mmHg) pulmonary hypertension.

### Statistical analysis

The Shapiro-Wilk test was used to determine if the data followed a normal distribution. For normally distributed data, the mean ± standard deviation was used, and t-tests were performed to compare differences between groups. For non-normally distributed data, the median and interquartile range were used, and the Mann-Whitney U test was performed to compare differences between groups. Count data were expressed as frequencies (percentages) and compared between groups using  $\chi^2$  tests or Fisher's exact tests. Univariate and multivariate logistic regression analyses were conducted to assess the association between various factors and adverse outcomes. All statistical analyses were performed using R 4.3.2 software, with  $\alpha=0.05$  as the test level and  $P<0.05$  considered statistically significant.

Variable	Overall		Univariate analysis for death		
	Death (n%)	Survival (n%)	OR	95% CI	P
Supracardiac	4 (28.6)	11 (32.4)			
Intracardiac	4 (28.6)	16 (47.1)	0.688	0.135–3.47	0.643
Infracardiac	4 (28.6)	6 (17.6)	1.833	0.326–10.62	0.486
Mixed	2 (14.2)	1 (2.9)	5.500	0.416–139.7	0.209
Pre-obstruction					
No	6 (42.9)	27 (79.4)			
Yes	8 (57.1)	7 (20.6)	5.143	1.37–20.94	0.017
CPV (cm) (mean (SD))	0.59 (0.19)	0.65 (0.18)	0.124	0.002–4.60	0.293
PFO (cm) (mean (SD))	0.55 (0.20)	0.57 (0.26)	0.693	0.030–8.95	0.792
LA (cm) (mean (SD))	0.80 (0.18)	1.02 (0.29)	0.012	0.000–0.290	0.015
PASP					
30–50 mmHg	1 (7.1)	10 (29.4)			
51–70 mmHg	8 (57.1)	13 (38.2)	6.154	0.906–124.0	0.111
> 70 mmHg	5 (35.7)	11 (32.4)	4.545	0.59–95.17	0.199
PDA (cm) (mean (SD))	0.57 (0.51)	0.44 (0.50)	1.689	0.484–6.16	0.414
Combined malformation					
No	11 (78.6)	23 (67.6)			
Yes	3 (21.4)	11 (32.4)	0.570	0.112–2.288	0.452
Sutureless					
No	13 (93)	28 (82.4)			
Yes	1 (7)	6 (17.6)	0.321	0.039–3.29	0.365
Years					
2014–2018	2 (14.3%)	10 (29.4)			
2019–2024	12 (85.7%)	24 (70.6)	0.770	-0.910–2.449	0.369
Age (day) (mean (SD))	34.79 (35.02)	128.71 (443.07)	0.992	0.974–1.001	0.281
Weight (kg) (mean (SD))	3.73 (0.94)	4.21 (1.94)	0.785	0.410–1.205	0.388

**Table 2.** Univariate logistic regression for postoperative death. *CPV* common pulmonary venous, *PFO* patent foramen ovale, *LA* left atrium, *PDA* patent duct arteriosus, *PH* pulmonary hypertension, *PASP* pulmonary arterial systolic pressure.

Variable	OR	95% CI	P
Pre-obstruction			
No			
Yes	3.399	0.745–15.497	0.114
LA (cm)			
≤ 0.85	ref		
> 0.85	0.143	0.024–0.857	0.033
Age (day)	0.998	0.989–1.008	0.759
Weight (kg)	1.807	0.626–5.219	0.274

**Table 3.** Multivariable logistic regression for postoperative death.

## Data availability

The data that support the findings of this study are available from the corresponding authors upon reasonable request.

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## Author contributions

Zongjie Weng designed and Shan Guo supervised the study. Xiaoying Xue worked on the literature review. Wen Ling focused on imaging acquisition, data collection, and analysis. Xiaoying Xue and Biying Huang performed the statistical analysis. Xiaoying Xue drafted the manuscript. Shan Guo worked on the editing. All authors contributed to the article and approved the submitted version.

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## Declarations

## Competing interests

The authors declare no competing interests.

## Additional information

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