



# Echocardiographic features and influencing factors of pulmonary hypertension of total anomalous pulmonary venous connection in adults

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**Background:** Patients with total anomalous pulmonary venous connection (TAPVC) generally have symptoms during the neonatal period and infancy, and the fatality rate is extremely high. Most patients do not survive to adulthood. This study analyzed the clinical and transthoracic echocardiographic (TTE) manifestations of adult patients with TAPVC, summarized the echocardiographic characteristics of TAPVC, and identified the factors influencing pulmonary hypertension.

**Methods:** Data from adult patients with TAPVC from Beijing Anzhen Hospital, China, were retrospectively collected for analyses, including sex, age, history of gestation, clinical manifestations, echocardiographic parameters, and blood oxygen levels. Patients were grouped for comparative analyses based on their pulmonary artery systolic pressure (PASP) ( $\geq 60$  vs.  $< 60$  mmHg); 32 atrial septal defect (ASD) patients were included as a control group.

**Results:** (I) Sixteen patients were identified with TAPVC (11 women and 5 men; mean age:  $32.2 \pm 9.5$  years), including 8, 4, and 4 patients with supra-cardiac, mixed, and intracardiac type TAPVC, respectively. Furthermore, 10 patients had moderate or severe tricuspid regurgitation, and 6 had a PASP of  $\geq 60$  mmHg. Echocardiography misdiagnosed 2 patients with an ASD. (II) The TAPVC group patients had a smaller left atrium (LA) and a lower aorta/pulmonary artery ratio than ASD-only group patients. However, the right ventricular diameter (RVd) and right atrium were larger in patients with TAPVC than in those with only ASD. (III) The RVd was larger and the LA was smaller in patients with a PASP of  $\geq 60$  mmHg than in those with a PASP of  $< 60$  mmHg. (IV) Of those with a PASP of  $\geq 60$  mmHg, TAPVC patients had a smaller LA and a larger RVd than those with only ASD. (V) Pregnancy affected the PASP (adjusted odds ratio: 15.000, 95% confidence interval: 1.031–218.300,  $P=0.047$ ). (VI) Echocardiography indicated that TAPVC patients with ASD had a right to left shunt at the atrial level and the pulmonary vein (PV) was not connected to the LA.

**Conclusions:** Searching for the PV by TTE is necessary for patients with ASDs, which may help avoid misdiagnosis. Moreover, pregnancy affects the PASP. Patients with TAPVC may present with a larger right heart, smaller LA, and lower aorta/pulmonary artery ration than those with only ASD.

**Keywords:** Total anomalous pulmonary venous connection (TAPVC); transthoracic echocardiography (TTE); congenital heart disease (CHD)

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## Introduction

Total anomalous pulmonary venous connection (TAPVC) accounts for 1.5–3.0% of all congenital heart diseases (CHDs) (1-4). Patients with TAPVC generally have symptoms, such as difficulty breathing, shortness of breath, and cyanosis, during the neonatal period and infancy. Moreover, for those with untreated TAPVC, 50% die within three months after birth, and increases to 80% die by the end of the first year after birth (5). In addition, increased pulmonary blood flow eventually leads to the muscularization of the pulmonary vascular bed, potentially causing irreversible pulmonary hypertension (PH). TAPVC can also lead to severe PH, which affects the treatment methods and prognosis. Thus, early detection and treatment are essential for optimal outcomes.

Patients with TAPVC rarely survive to adulthood, and the characteristics of TAPVC in adults remain unclear. Therefore, this study summarized the echocardiographic and clinical characteristics of TAPVC in adults and evaluated the factors affecting pulmonary artery systolic pressure (PASP) in these individuals. We present this article in accordance with the STROBE reporting checklist (available at <https://jtd.amegroups.com/article/view/10.21037/jtd-22-1793/rc>).

## Methods

### General data

Data from 16 adult patients with TAPVC diagnosed in Beijing Anzhen Hospital, China, between January 2002 to

December 2020 were retrospectively collected. Patients with TAPVC with atrial septal defects (ASDs) were included, but those with CHDs other than ASD were excluded. Data from 32 patients diagnosed only with ASD were also collected as a control group; the ASD diameters of those in the ASD-only group were matched with the ASD diameters of those in the TAPVC group.

### Anatomical features

TAPVC was classified as supra-cardiac, infracardiac, intracardiac, or mixed based on the drainage site of the abnormal veins as follows: (I) supra-cardiac type: the pulmonary vein (PV) was not connected to the left atrium (LA), forming a common cavity that drained into the innominate vein or superior vena cava through the vertical vein; (II) infracardiac type: a common cavity drained into the inferior vena cava or portal vein through the vertical vein; (III) intracardiac type: a common cavity converged with the coronary sinus or directly entered the right atrium (RA); and (IV) mixed type: the PV was connected to the RA or superior vena cava through different pathways.

### Ethics statement

The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the Ethics Committee of Beijing Anzhen Hospital (Approval No. 2022186X), and individual consent for this retrospective analysis was waived.

### Instruments and methods

Echocardiographic examinations were performed using a GE Vivid E9 ultrasound machine with an M5S probe (2–4 MHz; GE Healthcare, Chicago, IL, USA) or a Philips IE33 ultrasound machine with an S5-1 probe (2.5–3.5 MHz; Philips Healthcare, Andover, MA, USA).

### Data collection

General clinical data were collected, including height, weight, body surface area (BSA), clinical symptoms, medical history, and the blood oxygen (SpO<sub>2</sub>) level. Data on echocardiographic parameters were collected, including left ventricular end-diastolic diameter (LVEDd); LA anteroposterior, right ventricle (RV) transverse, aortic (AO), pulmonary artery (PA), maximum ASD, RA

### Highlight box

#### Key findings

- This report presents the clinical and transthoracic echocardiographic manifestations of 16 adult patients with TAPVC and demonstrates that pregnancy affects the PASP of adult patients with TAPVC.

#### What is known and what is new?

- Patients with TAPVC rarely survive to adulthood, and the characteristics of TAPVC in adults remain unclear.
- This study summarized the echocardiographic of TAPVC in adults and evaluated the factors affecting PASP in these individuals.

#### What is the implication, and what should change now?

- This study enriches the number of reports describing TAPVC patients in their adulthood, increasing the understanding of this disease.

**Table 1** Baseline data

Parameter	TAPVC	ASD	P
Age (years)	32.2±9.5	41.7±14.7	0.020*
Number of patients	16	32	–
TAPVC type			–
Supra-cardiac	8	–	
Intracardiac	4	–	
Mixed	4	–	
Sex (male/female)	5/11	16/16	0.175
Tricuspid regurgitation			–
Severe	7	7	
Moderate	3	10	
Slight	6	15	
PASP ≥60 mmHg	6	9	–
BSA (m <sup>2</sup> )	1.57±0.13	1.73±0.21	0.007*
Operation history	13	32	–
Pregnancy history (total)	8	12	–
SpO <sub>2</sub> (%)	88.6±4.2	97.3±1.3	<0.001*

Data are presented as mean ± standard deviation or number. \*, P<0.05. TAPVC, total anomalous pulmonary venous connection; ASD, atrial septal defect; PASP, pulmonary artery systolic pressure; BSA, body surface area; SpO<sub>2</sub>, blood oxygen level.

longitudinal, and RA transverse diameters. The AO/PA ratio and ASD/atrial septum ratio were also calculated. All echocardiographic parameters were standardized by the BSA. Furthermore, the PASPs of five patients was measured by right cardiac catheterization, and the PASP of 11 patients was estimated by tricuspid regurgitation (TR). If there was no obstruction of the RV outflow tract, the PASP was equal to the square of the TR velocity plus the RA pressure. The LVEDd and LA anteroposterior and AO diameters were measured on the left ventricular long-axis view. The RV basal segment transverse and RA diameter were measured on the apical four-chamber view. The PA diameter was measured on the short-axis view of the major artery.

### Patient groups

When the peak TR velocity was >3.4 m/s (PASP is ~60 mmHg), further intervention was needed (6). Thus, patients with TAPVC were divided into two groups: PASP <60 mmHg and PASP ≥60 mmHg; and patients with only ASD were accordingly divided into two groups: PASP <60 mmHg and PASP ≥60 mmHg.

### Statistical analyses

SPSS version 20.0 (IBM Corp., Armonk, NY, USA) was used for all statistical analyses. Quantitative variables were expressed as means ± standard deviations, and qualitative variables as percentages. *T*-tests were performed for classifying data, chi-square tests for counting data, and binary logistic regression for multivariate analyses. A *P* value of <0.05 was considered reflective of statistical significance.

## Results

### General data

This study included 32 patients with only ASD [16 women and 16 men; mean age: 41.7±14.7 (range, 18–73) years] and 16 adults with TAPVC [11 women and five men; mean age: 32.2±9.5 (range, 16–47) years] (Table 1). Of those with TAPVC, 10 patients had moderate or severe TR, and six had a PASP of ≥60 mmHg, five of whom were women, all with a history of pregnancy. Nine patients with only ASD had a PASP of ≥60 mmHg.

### Echocardiographic results

The AO inner diameter was significantly smaller than the PA diameter (Table 2).

Echocardiographic parameters were compared between the ASD-only and TAPVC groups. The LA was smaller and the AO/PA ratio was lower in the TAPVC group than in the ASD-only group. Additionally, patients with TAPVC had larger RVd and RAs and were younger than those with

**Table 2** Comparison table of the AO inner diameter and the PA diameter in the TAPVC groups

Group	Diameter (mm)
AO	25.91±5.22
PA	32.91±7.93
t	-2.445
P	0.023

Data are presented as mean ± standard deviation. AO, aorta; PA, pulmonary artery; TAPVC, total anomalous pulmonary venous connection.

only ASD (Tables 1,3).

### Effects of the PASP

Echocardiographic parameters were compared between the PASP ≥60 and <60 mmHg groups in patients with TAPVC. The LA was smaller and the RVd was larger in patients with TAPVC with a PASP ≥60 mmHg (Table 4).

Echocardiographic parameters were also compared between the ASD-only and TAPVC groups with a PASP of ≥60 mmHg. The RVd was larger and the LA was smaller in the TAPVC group than in the ASD-only group. The TAPVC group had more women than men, and the ASD-only group had more men than women (Table 5).

### Factors affecting the PASP in patients with TAPVC

Pregnancy, including a prior history of pregnancy or currently pregnant status, was a significant influencing factor of the PASP (adjusted odds ratio: 15.000, 95% confidence interval: 1.031–218.300, P=0.047) (Table 6).

**Table 3** Comparison table of the echocardiographic parameters in the ASD and TAPVC groups

Group	ASD (mm)	ASD/BSA (mm/m <sup>2</sup> )	ASD/AS ratio	LA/BSA (mm/m <sup>2</sup> )	LVEDd/BSA (mm/m <sup>2</sup> )	RV/BSA (mm/m <sup>2</sup> )	RA trans-D/BSA (mm/m <sup>2</sup> )	RA long-D/BSA (mm/m <sup>2</sup> )	AO/PA ratio
ASD	33.6±10.3	19.8±6.9	0.64±0.14	22.3±4.3	23.8±2.9	27.6±5.2	28.8±5.7	34.1±6.4	1.02±0.22
TAPVC	34.3±9.4	21.7±5.9	0.60±0.17	19.3±3.4	23.7±3.0	30.6±3.5	34.9±7.0	42.7±5.0	0.80±0.18
t	-0.230	-0.941	0.942	2.287	0.068	-2.019	-2.933	-4.015	3.241
P	0.751	0.351	0.351	0.027*	0.946	0.049*	0.005*	<0.001*	0.002*

Data are presented as mean ± standard deviation. \*, P<0.05. ASD, atrial septal defect; TAPVC, total anomalous pulmonary venous connection; BSA, body surface area; AS, atrial septum; LA, left atrium; LVEDd, left ventricular end-diastolic diameter; RV, right ventricle; RA trans-D, RA transverse diameter; RA long-D, RA longitudinal diameter; RA, right atrium; AO, aorta; PA, pulmonary artery.

**Table 4** The effects of PASP in the TAPVC group

Group	SpO <sub>2</sub> (%)	Age (years)	BSA (m <sup>2</sup> )	ASD/BSA (mm/m <sup>2</sup> )	ASD/AS ratio	LA/BSA (mm/m <sup>2</sup> )	LVEDd/BSA (mm/m <sup>2</sup> )	RV/BSA (mm/m <sup>2</sup> )	RA trans-D/BSA (mm/m <sup>2</sup> )	RA long-D/BSA (mm/m <sup>2</sup> )	AO/PA ratio
PASP ≥60 mmHg	88.1±3.09	32.0±4.9	1.60±0.07	21.7±5.4	0.58±0.18	17.3±2.9	22.1±2.7	33.8±3.0	36.6±5.5	45.1±2.6	0.76±0.18
PASP <60 mmHg	88.9±4.78	32.3±11.7	1.56±0.15	21.6±6.5	0.62±0.18	20.9±3.0	25.1±2.7	28.7±2.2	32.9±8.7	39.8±6.0	0.83±0.18
t	-0.308	-0.059	0.700	0.018	-0.348	-2.213	-2.012	3.938	0.856	1.951	-0.614
P	0.763	0.954	0.495	0.986	0.735	0.049*	0.069	0.001*	0.414	0.083	0.552

Data are presented as mean ± standard deviation. \*, P<0.05. PASP, pulmonary artery systolic pressure; TAPVC, total anomalous pulmonary venous connection; SpO<sub>2</sub>, blood oxygen level; BSA, body surface area; ASD, atrial septal defect; AS, atrial septum; LA, left atrium; LVEDd, left ventricular end-diastolic diameter; RV, right ventricle; RA trans-D, RA transverse diameter; RA long-D, RA longitudinal diameter; RA, right atrium; AO, aorta; PA, pulmonary artery.

**Table 5** Comparison of patients with PASP  $\geq 60$  mmHg in the ASD and TAPVC groups

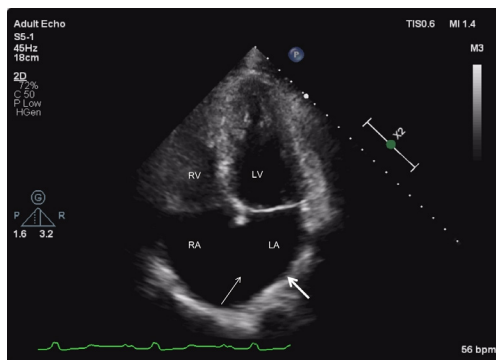
Group	Sex (male/female)	Age (years)	BSA (m <sup>2</sup> )	ASD/BSA (mm/m <sup>2</sup> )	ASD/AS ratio	LA/BSA (mm/m <sup>2</sup> )	LVEDd/BSA (mm/m <sup>2</sup> )	RV/BSA (mm/m <sup>2</sup> )	RA trans-D/BSA (mm/m <sup>2</sup> )	RA long-D/BSA (mm/m <sup>2</sup> )	AO/PA ratio
ASD	7/2	49.7 $\pm$ 12.2	1.73 $\pm$ 0.21	23.3 $\pm$ 4.0	0.71 $\pm$ 0.05	24.5 $\pm$ 3.4	23.5 $\pm$ 2.7	29.9 $\pm$ 2.7	33.8 $\pm$ 5.1	39.7 $\pm$ 5.7	0.92 $\pm$ 0.18
TAPVC	1/5	32.0 $\pm$ 4.9	1.60 $\pm$ 0.07	21.7 $\pm$ 5.4	0.58 $\pm$ 0.18	17.3 $\pm$ 2.9	22.1 $\pm$ 2.7	33.7 $\pm$ 3.0	36.6 $\pm$ 5.5	46.1 $\pm$ 2.6	0.76 $\pm$ 0.18
t	5.402	2.390	1.407	0.637	2.131	4.210	1.007	-2.542	-1.037	-2.083	1.614
P	0.020*	0.020*	0.183	0.535	0.053	0.001*	0.332	0.025*	0.318	0.059	0.133

Data are presented as mean  $\pm$  standard deviation. \*, P<0.05. PASP, pulmonary artery systolic pressure; ASD, atrial septal defect; TAPVC, total anomalous pulmonary venous connection; BSA, body surface area; AS, atrial septum; LA, left atrium; LVEDd, left ventricular end-diastolic diameter; RV, right ventricle; RA trans-D, RA transverse diameter; RA long-D, RA longitudinal diameter; RA, right atrium; AO, aorta; PA, pulmonary artery.

**Table 6** Binary logistic regression results for the factors influencing PASP

Variables	P	aOR	95% CI
Pregnancy	0.047	15.000	1.031–218.300
Age	0.607	0.972	0.872–0.083

PASP, pulmonary artery systolic pressure; aOR, adjusted odds ratio; CI, confidence interval.



**Figure 1** An apical four-chamber view of the heart showing the atrial septal defect (thin arrow) with a smooth left atrial wall (thick arrow). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; 2D, two-dimensional; TIS, thermal index of soft tissue; MI, mechanical index.

### Clinical manifestations

Eight patients with TAPVC recently had chest tightness, shortness of breath, and palpitations; of these, two had a PASP of  $\geq 60$  mmHg (68 and 101 mmHg with SpO<sub>2</sub> values of 83% and 96%, respectively). One patient had hemoptysis due to chest tightness (PASP: 106 mmHg; SpO<sub>2</sub>: 85.7%). One patient was post-menopausal and had a PASP of  $\geq 60$  mmHg (PASP: 80 mmHg; SpO<sub>2</sub>: 90%). Physical

examination indicated heart disease in one patient (SpO<sub>2</sub>: 93%). Furthermore, cyanosis occurred in five patients (SpO<sub>2</sub>: 84–88%) and PH in three patients (55, 68, and 90 mmHg).

### History of pregnancy, surgery, and treatment among patients with TAPVS

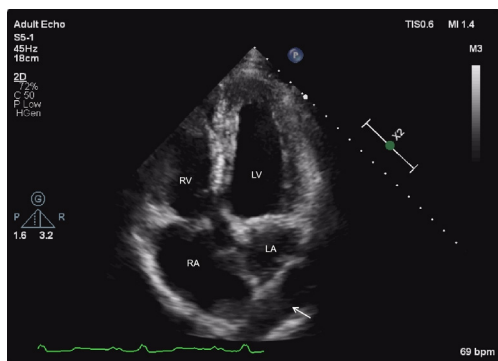
In total, 13 patients underwent repair operation, of which three patients were treated with medication for PH prior to surgical repair. Seven patients had a history of pregnancy and were 3–10 years postpartum, of which three patients were pregnant for a second or third time at the time of the diagnosis. One patient received surgical treatment, amongst other treatments (e.g., lowering PH), three months after terminating the pregnancy.

### Echocardiographic features

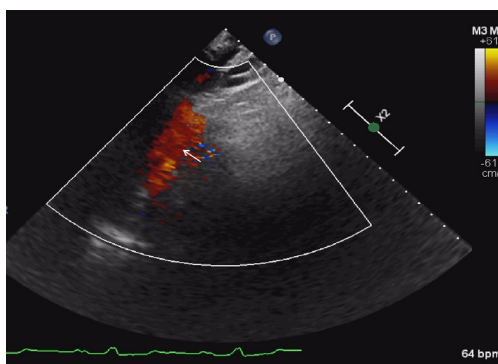
The echocardiographic features were as follows: (I) supra-cardiac type: the ASD and PV were not connected to the LA, forming a common cavity that drained into the innominate vein or superior vena cava through the vertical vein (*Figures 1-3*); (II) intracardiac type: the ASD and PV were not connected to the LA, forming a common cavity that converged with the coronary sinus or directly entered the RA (*Figure 4*); and (III) mixed type: the ASD and PV were not connected to the LA, and the PV was connected to the RA or superior vena cava through different pathways. No patients in this study had the infracardiac type.

### Misdiagnosis analysis

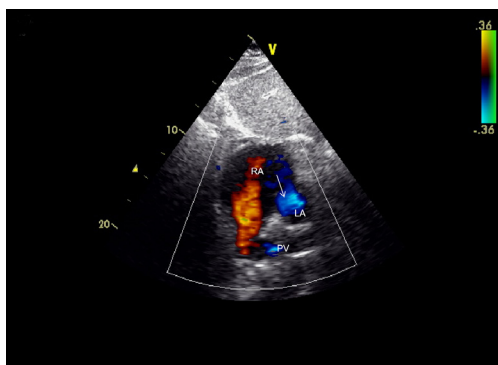
Two male patients were misdiagnosed. A large ASD was easily identified, but the connection between the PV and the common lumen structure was ignored. According to



**Figure 2** An apical four-chamber view of the heart showing the common lumen of pulmonary veins. The arrow indicates the common cavity. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; 2D, two-dimensional; TIS, thermal index of soft tissue; MI, mechanical index.



**Figure 3** CDFI: a suprasternal fossa view of the heart showing the vertical vein with an upward flow. The arrow shows the blood flow of the ascending vertical vein. CDFI, color Doppler flow imaging.



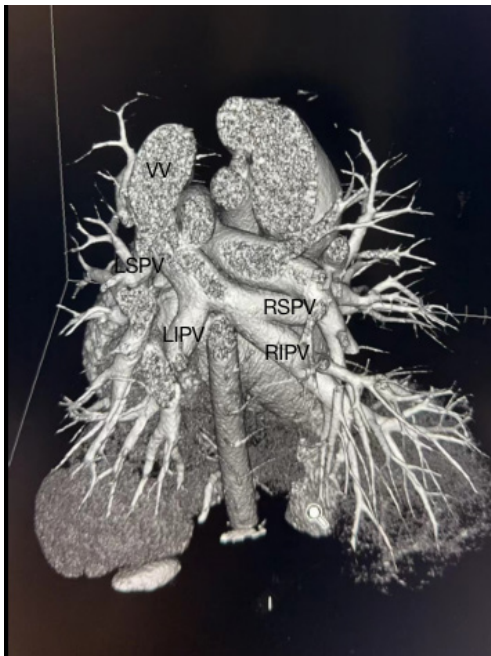
**Figure 4** An infra-xiphoid view of the heart showing that blood flows from the intracardiac PV into the RA and from the RA to the LA through the atrial septal defect (arrow). RA, right atrium; LA, left atrium; PV, pulmonary vein.

the analysis of the two patients, they were middle-aged men (44 and 47 years) with large ASDs, which explained the enlargement of the right heart. However, the doctor did not investigate the blood shunt direction at the ASD, nor locate the PV connection.

## Discussion

CHDs are common congenital disabilities and the primary cause of infant death. However, TAPVC is a rare cyanotic CHD characterized by an abnormal connection between the PV and the RA, which refluxes hyper-oxygenated blood into the right cardiac system. TAPVC is further classified as supra-cardiac, infracardiac, intracardiac, or mixed based on the drainage site of the abnormal veins. To survive TAPVC, patent foramen ovale or ASDs must exist. Furthermore, surgical intervention is required when the RV volume overloads and the patient experiences clinical symptoms and cyanosis. For those who survive to adulthood, mild clinical symptoms appear relatively late.

To survive to adulthood, patients with TAPVC must have no obstruction between the PV and systemic vein or RA, and the ASD size must be appropriate (4,5,7,8). Non-restrictive ASD provides suitable conditions for long-term survival. PV obstruction is more common in patients with infracardiac and mixed TAPVC, which can lead to severe pulmonary edema and clinical decompensation (9,10). Therefore, these types of TAPVC usually develop into severe pulmonary vascular disease early in life; thus, the patient will not survive to adulthood (5). However, no one in this study had infracardiac TAPVC nor PV obstruction. To our knowledge, the oldest patient with TAPVC is a 70-year-old woman who survived because she had a large ASD, no obstruction, and no related deformities and underwent successful TAPVC repair (11). In this study, the oldest patient was a 47-year-old man with supra-cardiac TAPVC locally diagnosed as ASD (maximum ASD diameter: 47 mm; slight TR) by physical examination. The supra-cardiac TAPVC was diagnosed by echocardiography in our hospital, and surgical treatment was performed with good postoperative outcomes. Autopsies of infants who died from TAPVC occlusion showed an increased thickness of the artery's middle layer, proliferation of the intima of the pre-sinus vein, and an abnormally small and thick wall of the external PV (12-14). This current study demonstrated that the cardiac structural changes in patients with TAPVC and only ASD are very similar; for example, as the right heart diameter increases, the left heart diameter



**Figure 5** Computed tomography angiography with supra-cardiac type TAPVC. RSPV, right superior pulmonary vein; RIPV, right inferior pulmonary vein; LSPV, left superior pulmonary vein; LIPV, left inferior pulmonary vein; VV, vertical vein; TAPVC, total anomalous pulmonary venous connection.

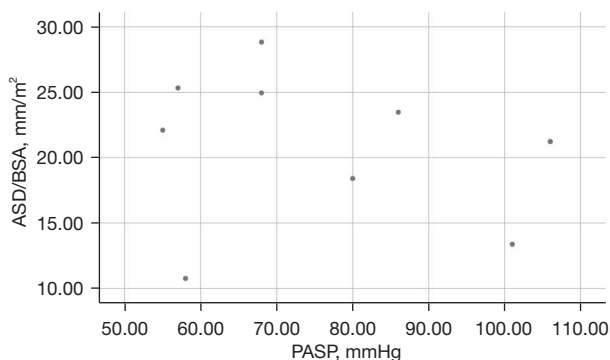
and AO/PA ratio decrease: this relationship is also the primary reason for missed diagnosis of TAPVC. Notably, the LA and AO/PA ratio were smaller and the RVD and RA were larger in patients with TAPVC. The hemodynamic changes of patients with TAPVC are more obvious than those of patients with only ASD. Patients with TAPVC have a wide range of clinical manifestations because of the vast differences in anatomy and hemodynamics, ranging from asymptomatic to severe hypoxemia; thus, TAPVC is most easily misdiagnosed as a larger, secondary ASD (15). These patients present with gradual symptoms, such as shortness of breath, atypical chest pain, or atrial arrhythmia (15-17). However, most TAPVC cases were diagnosed by echocardiography during the initial visit or during physical examination (18).

Two men in our cohort came to our hospital for surgery because of an ASD diagnosis, but supra-cardiac TAPVC was identified during the preoperative examination. There are a couple of explanations for missed diagnoses and misdiagnoses. First, the common chamber position is relatively biased, and abnormal structures cannot be

identified in the four-chamber view. Second, an ASD would explain the enlargement of the right heart; thus, if an ASD is present, the abnormalities of each structure are not always further explored, resulting in misdiagnosis. Transthoracic echocardiographic (TTE) is the preferred method for diagnosing isolated TAPVC; however, TTE may be considerably limited in its ability to assess PV abnormalities. Multidetector computed tomography (MDCT) (Figure 5) and magnetic resonance imaging (MRI) can also be used to accurately diagnose TAPVC. MDCT has 100% sensitivity and specificity for depicting the common PV drainage site, vertical vein stenosis, and atypical blood vessels entering the systemic veins. TTE also had 100% specificity for identifying the common PV drainage site, vertical vein stenosis, and atypical blood vessels entering the systemic veins, but the sensitivity of the common PV drainage site, vertical vein stenosis, and atypical blood vessels were 87%, 71%, and 0% respectively (18-20). Observing the direction of the blood shunt in the atrial septum and the PV connection is necessary if ASD is identified to avoid misdiagnosing TAPVC. However, when the specific return route is unclear, MDCT and MRI exams can provide more definite diagnostic information for the operation.

In this study, patients with TAPVC had a significantly smaller AO diameter than PA diameter, indicating that the increased volume of the right heart widened the PA. The RVD was larger and the LA was smaller in TAPVC patients with a PASP  $\geq 60$  mmHg than in those with a PASP  $< 60$  mmHg. Finally, of those with a PASP  $\geq 60$  mmHg, the RVD was larger and the LA was smaller in patients with TAPVC than in those with only ASD, and there were more women than men in the TAPVC group and more men than women in the ASD-only group. Pregnancy also significantly affected the PASP in patients with TAPVC. Maternal blood volume increases during pregnancy to meet the needs of the fetus; therefore, pregnancy dramatically affects the cardiovascular system, and the effects are sustained in the post-partum period (21). ASD size was not significant parameters related to PASP (Figure 6).

This study included a small number of patients, which is a limitation. However, the number of cases reported in the literature is also small because patients with TAPVC rarely live to adulthood. Notably, once TAPVC is diagnosed, it is surgically treated. Hence, relevant results on disease progression and changes cannot be obtained. Nevertheless, we plan to continue accumulating information on adult patients with TAPVC to study the related factors and disease changes of TAPVC-induced PH.



**Figure 6** Scatter diagram of PASP and ASD/BSA. PASP, pulmonary artery systolic pressure; BSA, body surface area; ASD, atrial septal defect.

## Conclusions

The structural changes in the heart of adult patients with TAPVC are similar to those of patients with ASD. Therefore, these two conditions must be differentiated based on the shunt at the atrial level and by identifying a common cavity. Furthermore, exploring the PV is very important for patients with ASD. Finally, pregnancy affected the PASP in patients with TAPVC, and the right heart was larger and the LA and the AO/PA ratios were smaller in patients with TAPVC than in those with only ASD.

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## Footnote

**Reporting Checklist:** The authors have completed the STROBE reporting checklist. Available at <https://jtd.amegroups.com/article/view/10.21037/jtd-22-1793/rc>

**Data Sharing Statement:** Available at <https://jtd.amegroups.com/article/view/10.21037/jtd-22-1793/dss>

**Peer Review File:** Available at <https://jtd.amegroups.com/article/view/10.21037/jtd-22-1793/prf>

**Conflicts of Interest:** All authors have completed the ICMJE uniform disclosure form (available at <https://jtd.amegroups.com/article/view/10.21037/jtd-22-1793/coif>). The authors have no conflicts of interest to declare.

**Ethical Statement:** The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the Ethics Committee of Beijing Anzhen Hospital (Approval No. 2022186X), and individual consent for this retrospective analysis was waived.

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