Congenital Heart Disease

Tech Science Press

DOI: 10.32604/chd.2022.018453

ARTICLE

Partial Anomalous Pulmonary Venous Connection and the Nature of Associated Sinus Venosus Defect

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Received: 26 July 2021 Accepted: 20 December 2021

ABSTRACT

Background: Partial anomalous pulmonary venous connection (PAPVC) is frequently associated with atrial septal defect (ASD), especially sinus venosus defect (SVD). Although Waggstaffe described the pathology of SVDs in 1868, the exact anatomic features and the nature of SVD remains controversial. SVDs with no posterior atrial rim were observed in recent years. However, no studies suggested that absence of the residual posterior atrial septal tissue might be the key feature of SVD. The aims of this study were to investigate if absence of posterior rim of atrial septum played a crucial role in patients with SVD. **Methods:** From January 2011 to December 2019, 256 children with PAPVC combined ASD and 878 children with isolated ASD who underwent corrective cardiac surgery were consecutively enrolled. Comprehensive review of preoperative transthoracic echocardiography, computed-tomography images and surgical findings were performed by experienced pediatric cardiologists. The subtypes of PAPVC, locations and types of ASD, and presence of posterior atrial rim of associated ASD were investigated. **Results:** PAPVC was right-sided in 244 children, left-sided in 6 children, and bilateral in 6 children. In PAPVC cases, ASD without posterior atrial rim existed in 226 SVD cases. ASD without posterior atrial septum only existed in cases with one or more right pulmonary veins returning to right atrium (RA) or to RA-superior vena cava junction. In cases with isolated ASD, there were 3 SVD, and the other 875 cases were secundum ASD. **Conclusions:** ASD without posterior atrial rims was associated with one or more right pulmonary veins returning to RA or RA-superior venous cava (SVC) junction. For SVD, the key feature is that the defect is in the posterior of the interatrial septum with no posterior septal rim, rather than adjacent to the SVC or to the inferior vena cava.

KEYWORDS

Partial anomalous pulmonary venous connection; sinus venosus atrial septal defect; echocardiography; right atrium; inferior vena cava

1 Introduction

Partial anomalous pulmonary venous connection (PAPVC) can involve right or left-sided pulmonary vein, which is drained to left innominate vein, hemiazygos vein, right atrium, coronary sinus and/or cava veins [1].

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Partial anomalous pulmonary veins are frequently associated with atrial septal defect (ASD), especially sinus venosus defect (SVD). It is estimated that approximately 80–90% of SVD cases have coexistent PAPVC [2,3], and SVD is a common form of interatrial communication in individuals with right pulmonary venous anomalous connection [1,4]. The association between PAPVC and SVD has been extensively studied in multidisciplinary teams, including pediatric cardiologists, pathologists and cardiac surgeons [1,5–7]. However, it remains unclear why SVD is commonly associated with right pulmonary venous anomalous connection, and the exact anatomical relationship between PAPVC and ASD is also undetermined.

The atrial septum has three components, namely septum primum, septum secundum and atrioventricular canal septum. ASD presents in isolated or multiple forms and locates any place within the atrial septum. ASD accounts for approximately 6–10% of congenital heart defect, with an incidence of 1 in 1,500 live births [4,8]. According to the anatomic location, ASD is classified into four types, namely secundum defect, primum defect, coronary sinus defect and SVD. SVD is a rare form of interatrial communication, which usually locates in superior or inferior part of the atrial septum, accounting for 11% of ASD [4,9–11]. It is well accepted that SVD is adjacent to superior vena cava (SVC) or inferior vena cava (IVC), and doesn't involve ossa ovale, septum primum and septum secundum. Although it is recognized that there are two types of SVD, namely superior and inferior SVD, the exact anatomic features and true nature of SVD remains unclear, and the accuracy of diagnosis is challenging [5,12,13]. Even with advancing imaging techniques, it remains difficult to differentiate SVD and secundum ASD with deficient superior and/or inferior rims [12]. In 2017, Snarr et al. reported that among patients with inferior SVD, all posterior rim was absent [11]. Absence of posterior rim was also observed in patients with superior SVD [5,10,14]. These findings together suggest that the absence of the posterior rim might play a crucial role in SVD.

In the current study, we reviewed individuals with PAPVC combined ASD that have underwent corrective cardiac surgery in the Guangdong Cardiovascular Institute, Guangdong Provincial People's Hospital, the largest tertiary cardiac center in China. The subtypes of PAPVC, location and types of associated ASD, presence of posterior rim of associated ASD, and features of SVD would also be discussed.

2 Methods

2.1 Study Participants

The retrospective study was approved by the Clinical Research Ethic Committee of Guangdong Provincial People's Hospital (the ethical approval number: GDREC 2016188H). From January 2011 to December 2019, 256 children with PAPVC combined ASD, that have undergone corrective cardiac surgery in Guangdong Provincial People's Hospital, were consecutively enrolled for the current study after parental written informed consent was obtained. Data of 878 consecutive children with isolated ASD, that have undergone surgical closure during the same period, were collected and served as the control group.

2.2 Data Collection

Transthoracic echocardiographic (TTE) examination was carried out preoperatively using IE33, IE Epic-7C (Philips, Andover, MA, USA) and Vivid E 9 (GE Healthcare, Horten, Norway) ultrasound systems with a transducer of 3–8 MHz or 1–5 MHz. And 256-slice scanner of multiple-detector computed tomography (MDCT; Brilliance iCT; Philips Healthcare, Cleveland, OH, USA) was performed preoperatively in 270 cases (n = 221 in patients with PAPVC combined ASD and n = 49 in patients with isolated ASD). The preoperative TTE images, MDCT images and surgical reports were reviewed by two independent experienced pediatric cardiologists (at least ten years' experience) to determine the location and subtype of ASD and the presence of posterior rim of ASD. If there were discrepancies between these two observers, another independent experienced pediatric cardiologist (at least fifteen years' experience) would be invited to review the images. The imaging data, including abnormal connected pulmonary vein, connection of abnormal pulmonary veins, location and subtype of ASD and presence of posterior atrial rim, was extracted and stored in the dataset.

2.3 Definition

In the present study, a superior SVD was defined as a defect located in superior and posterior region of the atrial septum and was adjacent to SVC [14,15]. An inferior SVD was defined as a defect located in inferior and posterior region of the atrial septum and was adjacent to IVC [12,15]. In the current study, besides superior and inferior SVD, there were two other special types of ASDs that were often associated with right sided PAPVC: these we called "posterior ASD" and "mixed SVD" for the purposes of this study. A posterior ASD was defined as an ASD in the posterior part of the atrial septum with no posterior atrial rim, which was neither adjacent to the SVC nor to the IVC. A mixed SVD was defined as an ASD with no residual posterior atrial septal tissue, which extended from the entrance of the SVC to the entrance of IVC. Secundum ASD was defined as a defect in central-most portion of the atrial septum, with extending to the surrounding septal part in some cases [4,14].

2.4 Statistical Analysis

Continuous variables were presented as mean ± standard deviation if normal distribution otherwise was presented as median (interquartile range). Between-group differences were analyzed using Student *t* test or Mann–Whitney U test as appropriate. Categorial variables were presented as number and proportion, and between-group differences were analyzed using Chi-squared test or Fisher exact test as appropriate. All analyses were performed using SPSS Statistics 23 (IBM, Chicago, IL, USA) and a two-sided *P*-value < 0.05 was considered as statistical significance.

3 Results

3.1 Clinical Characteristics

A total of 256 children with PAPVC combined ASD and 878 children with isolated ASD were included for the current analysis. The age of children with PAPVC combined ASD ranged from 0.08 to 14 years old, and 0.13 to 14 years old in children with isolated ASD. The body weight of children with PAPVC combined ASD ranged from 3.3 to 51 kg, and 2.3 to 84 kg in children with isolated ASD.

As presented in Table 1, compared to the isolated ASD group, children in the PAPVC combined ASD group were more likely to be boys, had a lower mean body weight at surgery, and were more likely to undergo preoperatively MDCT examination.

Table 1: Clinical characteristics

	PAPVC with ASD	Isolated ASD	*P*-value
n	256	878	
Male, n (%)	139 (54.3)	397 (45.2)	0.013
Body weight at surgery (kg)	13.5 ± 8.2	16.0 ± 11.0	<0.0001
Age at surgery (months)[a]	35.5 (12–63)	36.0 (17–78)	0.29
MDCT, n (%)	211 (82.4)	49 (5.6)	<0.0001

Note: MDCT, Multiple detector computed tomog raphy; [a]Presented as median (interquartile).

3.2 Adjacent Anatomy of Atrial Septal Defects in Surgery Founding

Types of ASD and adjacent anatomical relationships were summarized in Tables 2 and 3. All the patients undergone the surgery procedure. According to the surgery anotomy founding, SVD can be divided to superior SVD and inferior SVD. Apart from, two special types of ASD can be found, that were posterior ASD and mixed SVD. The anatomy character of posterior ASD was that ASD was located in the posterior part of the atrial septum with no posterior atrial rim, which was neither adjacent to the SVC nor to the IVC. The anatomy character of mixed SVD was that ASD was very large, extended from the entrance of the SVC to the entrance of IVC and no residual posterior atrial septal tissue.

Table 2: Types of PAPVC and associated ASD

Types of PAPVC	n	Superior SVD	Inferior SVD	Posterior ASD	Mixed SVD	Secundum ASD
Right-sided PAPVC	**244**	**65**	**127**	**31**	**2**	**19**
RSPV (RSPV+RMPV) to proximal SVC (SVC-RA junction)	38	35	3			
RSPV to distal SVC+RMPV to proximal SVC (SVC-RA junction)	2	2				
RSPV (RSPV+RMPV) to distal SVC	13					13
RSPV (RSPV+RMPV) to RA	21	15	5	1		
All RPV to RA	107	13	69	23	2	
All RPV to IVC	4					4
All RPV-CS-RA	2					2
RIPV (RIPV+RMPV) to RA	57		50	7		
Left-sided PAPVC	**6**					**6**
Bilateral PAPVC	**6**		**1**			**5**
All RPV to RA+LSPV to Inn. vein	1		1			
LPV+RLPV (RLPV+RMPV)-CS-RA	2					2
LPV+RSPV to Inn. vein	1					1
RSPV (RSPV+RMPV) to distal SVC +LSPV to Inn. vein	2					2
Total	**256 (100%)**	**65 (25.4%)**	**128 (50.0%)**	**31 (12.1%)**	**2 (0.8%)**	**30(11.7%)**

Note: ASD, atrial septal defect; Inn. Vein, innominate vein; IVC, inferior vena cava; LPV, left pulmonary vein; LSPV, left superior pulmonary vein; PAPVC, Partial anomalous pulmonary venous connection; RA, right atrium; RIPV, right inferior pulmonary artery; RMPV, right middle pulmonary vein; RPV, right pulmonary vein; RSPV, right superior pulmonary vein; SVC, superior vena cava; SVD, sinus venosus defect.

Table 3: Types of ASD and adjacent anatomy

Types of ASD	Adjacent to the SVC	Adjacent to the IVC	Adjacent to the posterior atrial wall
Superior SVD	Yes	No	Yes
Inferior SVD	No	Yes	Yes
Posterior ASD	No	No	Yes
Mixed SVD	Yes	Yes	Yes
Secundum ASD	No	No	No

3.3 Types of PAPVC and Associated ASD

Types of PAPVC and associated ASD were summarized in Table 2. The proportion of right-sided, left-sided and bilateral PAPVC were 95.31% (n = 244), 2.34% (n = 6) and 2.34% (n = 6), respectively. Among 256 children with PAPVC combined ASD, 25.4% (n = 65), 50.0% (n = 128), 12.1% (n = 31), 0.8% (n = 2) and 11.7% (n = 30) had superior SVD (Fig. 1), inferior SVD (Fig. 2), posterior ASD (Figs.

3 and 4), mixed SVD and secundum ASD (Figs. 5 and 6), respectively. Interesting, except secundum ASD, other types of ASD combined no left sided PAPVCs. And superior SVD only combined right-sided PAPVC, inferior SVD can combined bilateral PAPVC, not only combined right-sided PAPVC easily, but also combined left-sided PAPVC.

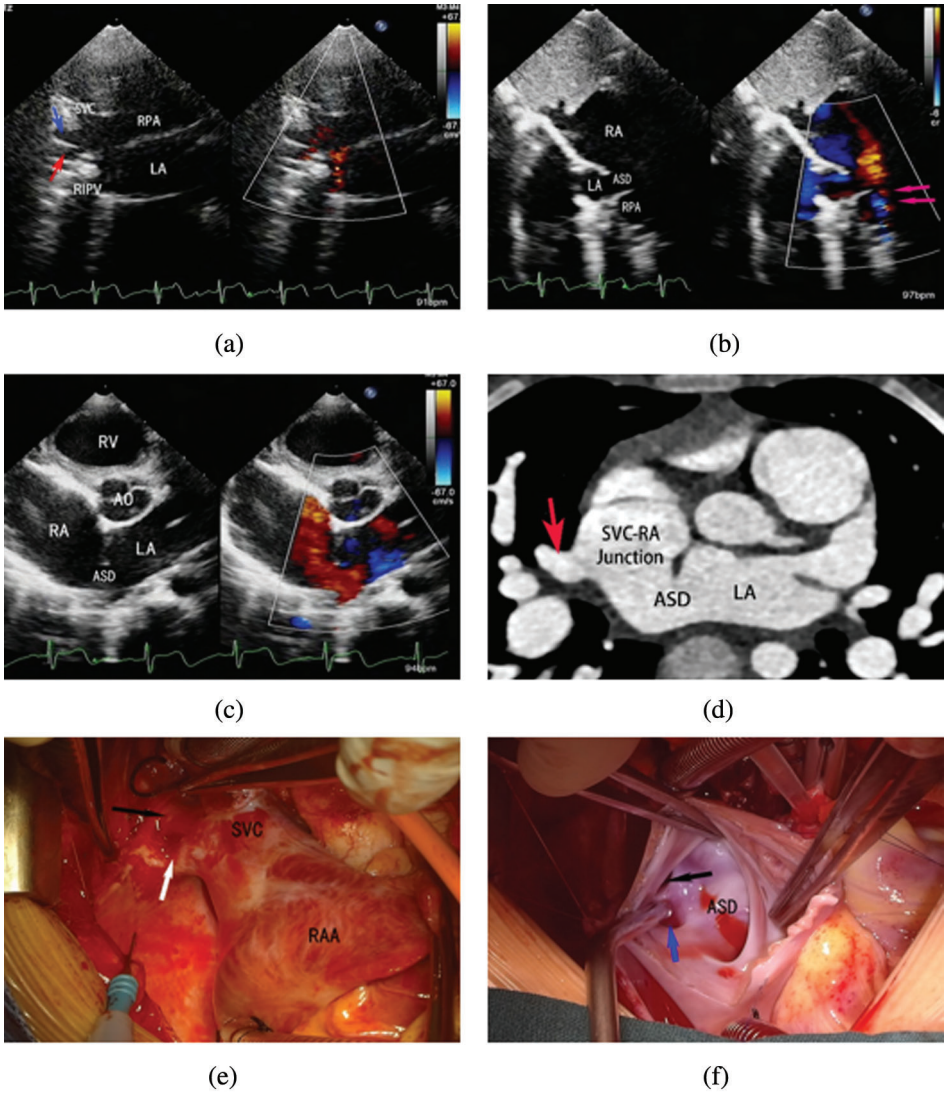


Figure 1: Right-sided PAPVC and associated superior SVD. (a) Supra-sternal echocardiographic view. RSPV (blue arrow) and RMPV (red arrow) connecting to SVC-RA junction. (b) Subcostal sagittal echocardiographic view shows a superior SVD/ASD. Arrow indicates SVC. (c) Parasternal short axis echocardiographic view. There was no posterior interatrial septal rim between ASD and posterior atrial wall. (d) Multiplanar reconstruction of multidetector CT showed RSPV connecting to SVC-RA junction. Absence of posterior rim of ASD was also observed. (e, f) Opened-up RA showed RSPV (black arrow) and RMPV (blue arrow) returning abnormally. An ASD in superior and posterior part of atrial septum with no residual posterior atrial septal tissue. (PAPVC, partial anomalous pulmonary venous connection; SVD, RSPV, right-sided pulmonary vein; RMPV, right middle pulmonary vein; SVD, sinus venosus defect; SVC, superior venous cava; RA, right atrium; ASD, atrial septal defect)

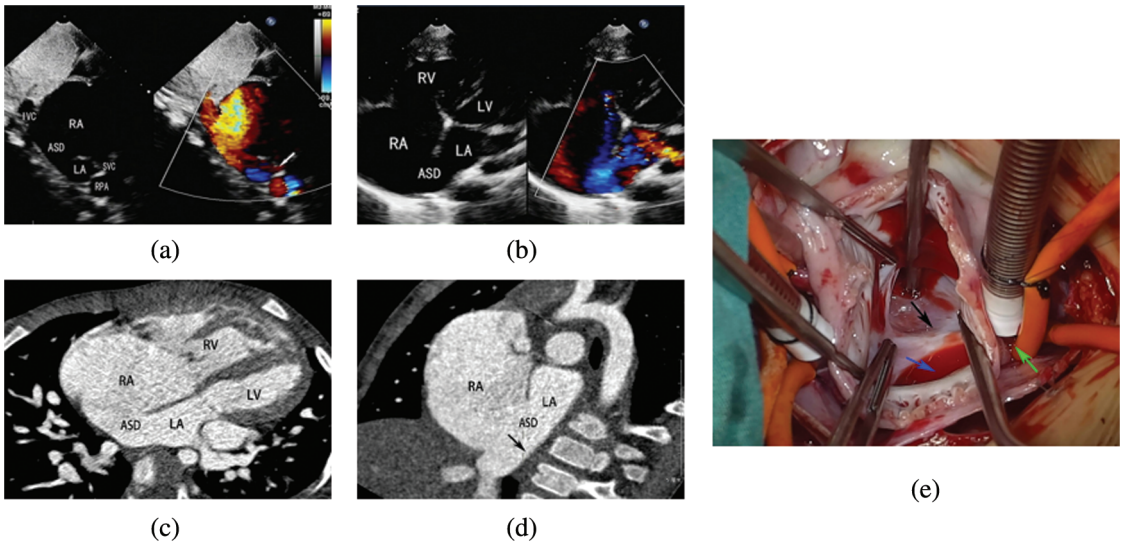


Figure 2: Inferior SVD with no posterior atrial rim and right pulmonary vein connected to RA. (a) Subcostal sagittal echocardiographic view demonstrated atrial septal defect adjacent to inferior vena cava. (b) Apical four-chamber view. ASD was in posterior region of atrial septum, with no residual posterior atrial septal tissue. (c) There was no posterior interatrial septal rim between ASD and posterior atrial wall in CT imaging. (d) Multiple-planar reconstruction CT demonstrated ASD without posterior atrial rim was adjacent to inferior vena cava. (e) Surgical findings. Opened-up RA showed that an inferior SVD was adjacent to inferior vena cava (green arrow demonstrates inferior vena cava cannula), with a 'bald' posterior wall (black arrow). Blue arrow indicated residual blood that was originated from anomalous right pulmonary veins. (SVD, sinus venosus defect; RA, right atrium; ASD, atrial septal defect)

3.4 Types of Drainage Location and Associated ASD Except Secundum ASD

Types of drainage location and associated ASD except secundum ASD were summarized in Table 4. Among 226 cases with PAPVC combined ASD, 17.7% (n = 40), 82.3% (n = 186), 0 (n = 0) connected to SVC, RA and IVC, respectively. Most of anomalous pulmonary venous connected to SVC in superior SVD (56.92%, n = 37), while connected to RA in inferior SVD (97.66%, n = 125). In posterior ASD and mixed SVD, anomalous pulmonary venous both connected to RA.

3.5 Posterior Atrial Rim in Cases with PAPVC Combined ASD

As shown in Tables 5 and 6, among 256 cases with PAPVC combined ASD, 226 (88.3%) did not have posterior atrial rim and all these cases were right-sided PAPVC, which were characterized by most of right pulmonary veins were drained to RA or RA-SVC junction. The common types of PAPVCs were all anomalous connected right pulmonary veins (47.35%, n = 107), second were anomalous connected right superior pulmonary vein and right middle pulmonary vein (26.99%, n = 61) in without posterior rim cases.

Among 244 cases with right-sided PAPVC combined ASD, 225 (92.2%) did not have posterior atrial rim, and 225 cases had one or more right pulmonary veins were drained to RA or RA-SVC junction. 19 cases (7.79%) had posterior atrial rim, including cases with right pulmonary veins connected to coronary sinus, and right pulmonary veins connected to distal part of SVC or IVC, respectively.

As shown in Table 7, in cases with isolated ASD, 2 cases had inferior SVD, 1 case had superior SVD, and the other 875 cases had secundum ASD. Posterior rim of ASD was observed in all cases except for cases with superior and inferior SVD. There were differences in the proportion of cases without posterior atrial rim between the PAPVC combined ASD and isolated ASD groups.

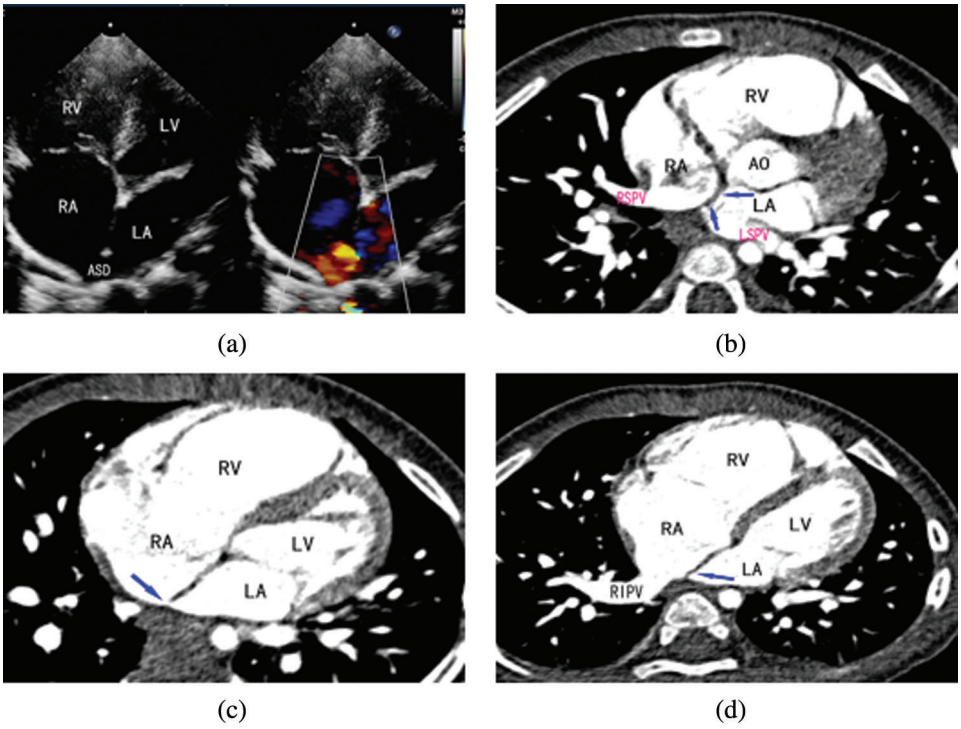


Figure 3: Right pulmonary veins connecting to RA and posterior ASD in an 8-year-old boy. (a) Apical four chamber view showed a posterior ASD. (b) CT section at the level of right superior pulmonary vein and left superior pulmonary vein. Right superior pulmonary vein connected to RA. Atrial septum was intact at this level. (c) Cross-sectional image beneath right superior pulmonary vein level. There was a posterior ASD above right inferior pulmonary vein. (d) CT at right inferior pulmonary vein level. Atrial septum was also intact in this section. It was obvious that this type of ASD was neither adjacent to superior vena cava nor to inferior vena cava (RA, right atrium; ASD, atrial septal defect)

4 Discussion

Partial anomalous pulmonary venous connection exhibits a wide anatomic spectrum. Almost every conceivable connection between pulmonary veins and systemic venous tributaries can be presented [3]. Right pulmonary vein draining to the cardiac end of right-sided SVC or right atrium is often associated with SVD. While SVD is often absent in cases with right-sided pulmonary vein (RSPV) connecting to distal part of SVC or with a left-sided PAPVC [14], which is consistent to the current findings. In the current study, in cases with RSPV connecting to distal part of SVC or with a left-side PAPVC, there was no inferior SVD, superior SVD, posterior ASD, or mixed SVD. Since SVD is absent in individuals with RSPV connecting to distal part of SVC, pediatric cardiologists and sonographers rarely focus on RSPV and SVC during echocardiographic examination. In addition, pulmonary venous flow is inconspicuous in acoustic window, therefore, RSPV connecting to distal part of SVC might be missed easily using TTE.

Scimitar syndrome is a special type of PAPVC, which refers to anomalous right pulmonary vein draining into IVC along with hypoplasia of right lung, secondary dextrocardia, and arterial supply of partial right lung by collateral vessels. It remains undetermined that whether Scimitar syndrome is commonly associated with SVD. It is reported that an interatrial communication is not commonly found in adults with Scimitar syndrome [16]. However, some studies have shown that SVD can present in cases with right pulmonary vein connecting to IVC. In the current study, no SVD was found in Scimitar syndrome.

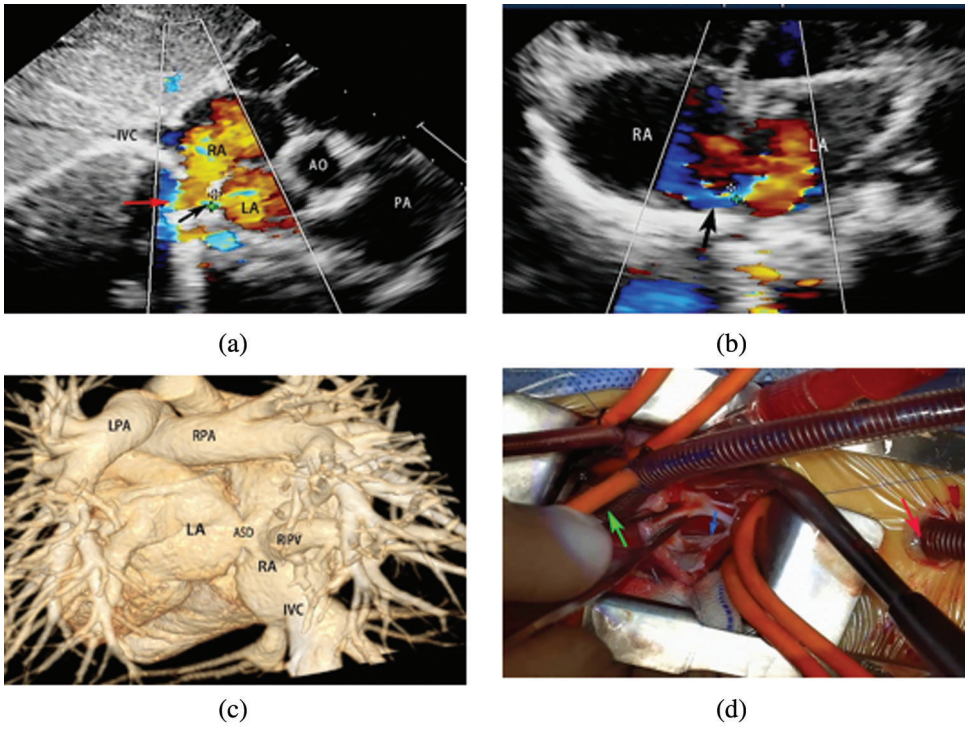


Figure 4: Posterior ASD in a 4-year-old boy with right pulmonary veins connected to RA. (a) Subcostal sagittal view demonstrated that right inferior pulmonary vein connected to RA with a posterior ASD. There was no posterior rim between ASD and posterior atrial wall. Red arrow indicates right pulmonary vein, black arrow indicates posterior ASD. (b) Apical four chamber view of posterior ASD. Black arrow indicates posterior ASD. (c) Volume-rendered CT image of right pulmonary veins anomalous connection with a posterior SVD. ASD was neither adjacent to SVC nor to inferior vena cava. (d) Opened-up RA showed that posterior ASD without posterior rim (blue arrow) was neither adjacent to superior vena cava nor to inferior vena cava. Red arrow indicates inferior vena cava cannula and green arrow indicates superior vena cava cannula (ASD, atrial septal defect; RA, right atrium; SVD, sinus venosus defect)

Prior studies have shown that most anomalous pulmonary veins arise from right side and primarily connect to SVC, and less commonly connect to RA or IVC [9,17,18]. Ammash et al. reported 45 cases with PAPVC, with 81.4% were right-sided, 16.3% left-sided, and 2.3% bilateral veins [17]. They found that the most common type of anomalous pulmonary vein was right superior pulmonary vein, with 48.5% of the cases, and most drained to SVC, with 59.1% of the cases. Importantly, the current findings are somehow different from prior reports [9,17,18]. In the present study, we enrolled 256 children with PAPVC combined ASD who underwent corrective cardiac surgery, and to the best of our knowledge, this should be the largest study of PAPVC and associated ASD. The findings demonstrate that the most common anomalous pulmonary vein was right pulmonary vein that commonly returned to RA and the most common type of SVD was the inferior type.

The pathology of SVD was first described by Waggstaffe in 1868 and once named high ASD as only superior SVD was recognized in the first decades [19]. In 1956, Ross et al. coined the term “sinus venosus defect” when he defined this defect as incomplete absorption of sinus venosus into right atrium [20]. With the advancement of cardiac surgery, the number of studies about SVD and PAPVC were increasing gradually. Nonetheless, up to now, the exact definition and nature of SVD remains controversial, and there are two widely recognized viewpoints. One is that the nature of SVD is

"unroofing" of pulmonary veins, and the other is that communications outside the confines of true septum. In 1994, van Praagh et al. reported that SVD appeared to have a deficiency in the wall that separates right pulmonary veins from right sinus venosus, and considered that one of the diagnostic criteria of SVD was actually "unroofing" of pulmonary veins [6]. They insisted that the interatrial communication was not a real defect. It is the left atrial orifice of the unroofed pulmonary veins as in cases of an unroofed coronary sinus (CS) where the interatrial communication is not a defect, rather, it represents the right atrial orifice of the CS. This theory can explain why superior SVD is commonly accompanied by anomalous connection of right superior pulmonary vein and why inferior SVD is often accompanied by anomalous connection of right inferior pulmonary vein. Nonetheless, this theory is challenged by other scholars. In the current study, most inferior SVD cases had right inferior and/or right superior pulmonary veins which directly connected to right atrium from posterior-lateral part, and there was no space to form a roof between right pulmonary veins and left atrium. In addition, right inferior pulmonary vein connecting to right atrium at the point near the mouth of IVC existed in superior SVD cases. Inferior SVD cases with right superior pulmonary vein connecting to SVC-RA junction also observed in the current study. Notably, the unroofing theory could not explain these cases. Considering these reasons, the idea of "unroofing" of right pulmonary veins seems questionable. SVD allows an atrial level or a supra-atrial level shunt. However, many scholars insisted that SVD was characterized by the defect being outside the confines of the true atrial septum and they were not true atrial septal defects [21,22]. McCarthy et al. defined a true septal structure as one that can be removed without transgressing on pericardial cavity [21]. It was believed that such communication outside the confines of the true septum was the key to differentiate SVD from true atrial septal defects (secundum ASD) [22,23]. However, the true interatrial septum is a relatively small area occupying about 20% of the whole atrial septum area [24]. In clinical practice, large secundum ASD is common, and these defects inevitably involve the outside tissues of the small true septum. Furthermore, the secundum ASD extending to the inferior and/or posterior part can also involve the outside tissues of the true septum as observed in the current and prior study [12]. With these theories, communications outside the confines of the true septum should not considered as the nature of SVD.

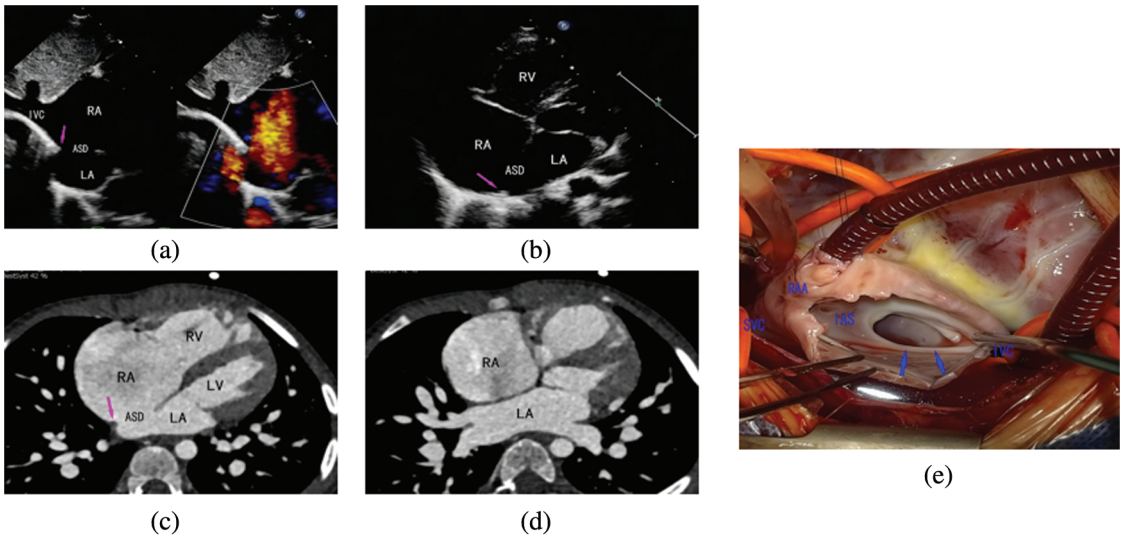
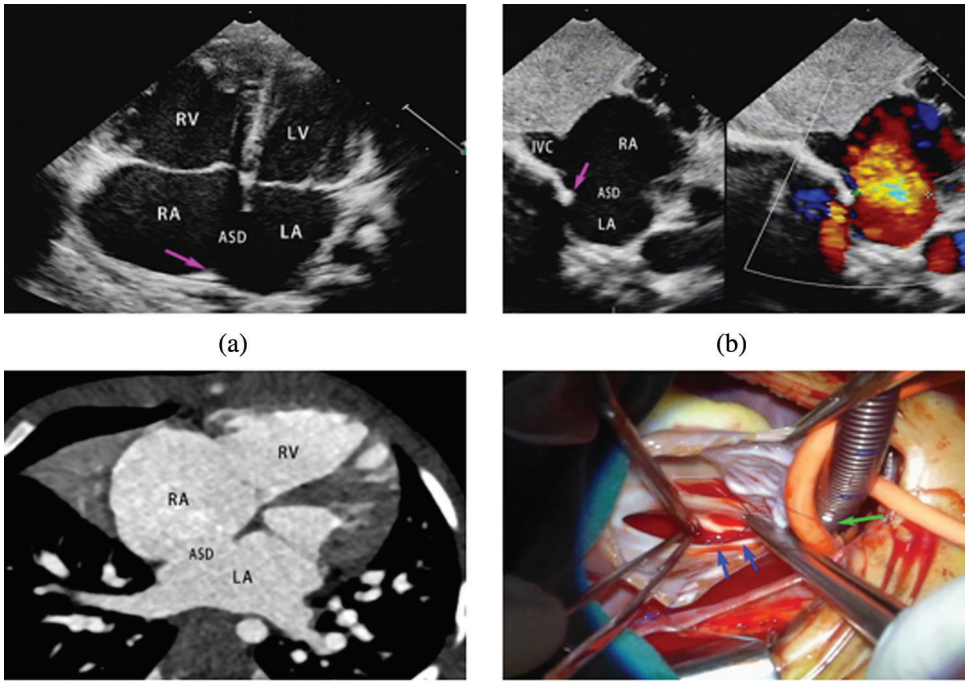


Figure 5: Images of "secundum ASD" adjacent to inferior vena cava. (a) Echocardiographic subcostal sagittal view demonstrated that secundum ASD was adjacent to inferior vena cava, but a posterior and inferior atrial septal rim (arrow) was present. (b) Apical four-chamber echocardiographic view. ASD and posterior atrial septal rim (arrow) were shown. (c) Posterior atrial septal rim was also detected by CT. (d) Right and left pulmonary veins returned normally. (e) ASD and posterior atrial septal rim (arrow) (ASD, atrial septal defect)



(a) (b)

(c) (d)

Figure 6: Large secundum ASD adjacent to inferior vena cava. (a) Large secundum ASD with a residual posterior atrial septal rim (arrow) were shown in apical four-chamber view. (b) Subcostal sagittal view. ASD was adjacent to inferior vena cava with a posterior and inferior atrial septal rim (arrow) were shown. (c) CT image. Large ASD and pulmonary veins returned normally. (d) Surgical findings demonstrated that an oval shaped large ASD was adjacent to inferior vena cava. Green arrow indicates inferior vena cava cannula. Blue arrow indicates posterior atrial rim of ASD (ASD, atrial septal defect)

Table 4: Types of drainage location and associated ASD

Drainage location	N (%)	Superior SVD	Inferior SVD	Posterior ASD	Mixed SVD
SVC-RA junction	40 (17.70)	37 (56.92)	3 (2.34)	0	0
RA	186 (82.30)	28 (43.08)	125 (97.66)	31 (100.00)	2 (100.00)
IVC	0	0	0	0	0
Total	226 (100.00)	65 (28.76)	128 (56.64)	31 (13.72)	2 (8.80)

Table 5: Cases with and without posterior atrial rim in the overall PAPVC and right-sided PAPVC

	n (%)	ASD	
		Without posterior rim	With posterior rim
Overall PAPVC with ASD	**256**	**226**	**30**
One or more RPVs to RA or to RA-SVC junction	226 (88.28)	226 (100.00)	0 (0.00)
Other type of PAPVC	30 (11.72)	0 (0.00)	30 (100.00)

(Continued)

Table 5 (continued)

	n (%)	ASD	
		Without posterior rim	With posterior rim
Right-sided PAPVC with ASD	**244**	**225**	**19**
One or more RPVs to RA or to RA-SVC junction	225 (92.21)	225 (100.00)	0 (0.0)
All RPV to IVC	4 (1.64)	0 (0.00)	4 (21.05)
RPV-CS-RA	2 (0.82)	0 (0.00)	2 (10.53)
RSPV (RSPV+RMPV) to distal SVC	13 (5.33)	0 (0.00)	13 (68.42)

Note: ASD, atrial septal defect; Inn. Vein, innominate vein; IVC, inferior vena cava; LPV, left pulmonary vein; LSPV, left superior pulmonary vein; PAPVC, Partial anomalous pulmonary venous connection; RA, right atrium; RIPV, right inferior pulmonary artery; RMPV, right middle pulmonary vein; RPV, right pulmonary vein; RSPV, right superior pulmonary vein; SVC, superior vena cava; SVD, sinus venosus defect.

Table 6: Cases with and without posterior atrial rim in the different types of PAPVC

	n (%)	Without posterior rim	With posterior rim
RSPV+RMPV	74 (28.91)	61 (26.99)	13 (43.33)
All RPV	113 (44.14)	107 (47.35)	6 (20.00)
RIPV+RMPV	57 (22.27)	57 (25.22)	0 (0)
LPV	6 (2.34)	0 (0)	6 (20.00)
RPV+LPV	6 (2.34)	1 (0.44)	5 (16.67)
Total	256	226	30

Note: LPV, left pulmonary vein; RIPV, right inferior pulmonary artery; RMPV, right middle pulmonary vein; RPV, right pulmonary vein; RSPV, right superior pulmonary vein.

Table 7: Types of ASDs in PAPVC combined ASD and isolated ASD groups

	PAPVC combined ASD	Isolated ASD	Total	*P*-value
Superior SVD, n (%)	65 (25.4)	1 (0.1)	66	
Inferior SVD, n (%)	128 (50.0)	2 (0.2)	130	
Posterior ASD, n (%)	31 (12.1)	0	33	
Mixed SVD, n (%)	2 (0.8)	0	0	
Secundum ASD, n (%)	30 (11.7)	875 (99.7)	905	
Total	256	878	1134	<0.001

It is well known that there are two types of SVD, with one is superior SVD and the other is inferior SVD [4,5,7,25]. The superior SVD located in superior and posterior region of atrial septum and is adjacent to SVC, and the inferior SVD is located in inferior and posterior region of atrial septum and is adjacent to IVC. There are different explanations about the nature of SVD, and SVD locates in posterior part of atrial septum is often mentioned [5,11,14,25,26]. However, surgeons and sonographers commonly pay close attention to SVDs being superior or inferior, as to the residual posterior atrial septal tissue of SVD exited or not, it was rarely turned into a focus problem. In the present study, we found that all SVD, regardless of superior or inferior, located in posterior part of atrial septum with no septal tissue separating the defect from posterior atrial wall, exhibiting a 'bald' appearance to posterior atrial wall. In 2017, Snarr et al. [11] first described

the “bald posterior wall” theory. Based on results of TTE examination from 15 inferior SVD patients and 14 secundum ASD patients, they found that the posterior rim was absent in all SVD patients while was present in all 14 patients with secundum ASD. In the current study, there were 31 cases with posterior ASD (the defect is neither adjacent to the SVC, nor to the IVC). Similar to superior SVD and inferior SVD, posterior ASD has no residual atrial septal tissue in posterior atrial wall and is commonly associated with abnormal right pulmonary vein connection. Indeed, posterior ASD without posterior rim has been reported as one type of SVD. In 2017, Tretter et al. [13] performed an autopsy in a special SVD case. The findings showed that the small defect was neither adjacent to IVC nor to SVC, and it just located in posterior part with a “bald posterior atrial wall”. Other scholars called this type of atrial communication as an atypical SVD [9]. In the current study, there was another special type of ASD that was associated with right pulmonary vein connecting to RA, which we called mixed SVD. This oval-shaped atrial communication extended from the mouth of SVC and connected to the mouth of IVC. There was no posterior atrial septal rim, and these detects were adjacent to both SVC and IVC. While in the control group, other than 3 SVD cases, there were no case of ASD with “bald posterior wall”.

It was clearly that, whatever superior SVD, inferior SVD, posterior ASD and mixed SVD, all these defects have the common features, that is absent of the posterior atrial rim and almost all are associated with one or more right pulmonary veins connecting to the RA or to the SVC-RA junction. Based on these findings, we propose that ASD without posterior septal rim should be identified as SVD, which should be considered the exact nature of SVD. During week 5 of gestation, a plexus of veins that envelope the bronchial buds meet with developing pulmonary veins so as to establish a connection. As the left atrium develops, it progressively incorporates the common pulmonary vein into the left atrial wall until all four pulmonary veins enter the posterior wall of the left atrium separately. The incorporated pulmonary veins form the smooth posterior wall of the left atrium, whereas the trabeculated portion of the left atrium occupies a more ventral aspect [24,27]. In healthy individuals, right pulmonary veins are close to the posterior interatrial septum. Hence, in the embryo stage, developmental disorder of the posterior part of atrial septum and adjacent right pulmonary vein will lead to SVD and a right-sided anomalous pulmonary venous connection. This hypothesis could explain why: 1) SVD is often accompanied with right-sided anomalous pulmonary venous connection; 2) SVD is often absent in patients with the RSPV connecting to distal parts of the SVC and in patients with a left-sided PAPVC. In these patients, the anomalous pulmonary veins are far from the atrial septum, therefore, malformation of pulmonary veins will not affect development of the interatrial septum.

5 Conclusion

In conclusion, ASD without posterior atrial rims was associated with one or more right pulmonary veins returning to RA or RA-superior venous cava (SVC) junction. For SVD, the key feature is that the defect is in the posterior of the interatrial septum with no posterior septal rim, rather than adjacent to the SVC or the inferior vena cava.

Acknowledgement: We appreciate very much for all participants and their family. We also thank very much for Dr. Anping Cai who has critically revised this paper.

Authors’ Contribution: Ling Sun, Chengcheng Pang, Xiaoyan Wang: Conceptualization, Methodology, Software, Investigation, Formal analysis, Writing—Original draft; Ming-guo Xu: Resources, Data curation; Shushui Wang and Zhiwei Zhang (Corresponding author): Conceptualization, Writing—Review & Editing, Supervision, Funding acquisition.

Funding Statement: The current study was supported by the National Key R&D Program of China [2018YFC1002600], Science and Technology Planning Project of Guangdong Province [Nos. 2018B090944002,

2019B020230003 and 2018KJY2017], Guangdong Peak Project of Guangdong Province [DFJH201802] and the Shenzhen San-Ming Project.

Conflicts of Interest: The authors declare that they have no conflicts of interest to report regarding the present study.

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