Cardiology in the Young

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Multimodality imaging in delineation of complex sinus venosus defects and treatment outcomes over the last decade*

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Abstract

Background: Diagnosis of sinus venosus defects, not infrequently associated with complex anomalous pulmonary venous drainage, may be delayed requiring multimodality imaging. Methods: Retrospective review of all patients from February 2008 to January 2019. Results: Thirty-seven children were diagnosed at a median age of 4.2 years (range 0.5–15.5 years). In 32 of 37 (86%) patients, diagnosis was achieved on transthoracic echocardiography, but five patients (14%) had complex variants (four had high insertion of anomalous vein into the superior caval vein and three had multiple anomalous veins draining to different sites, two of whom had drainage of one vein into the high superior caval vein). In these five patients, the final diagnosis was achieved by multimodality imaging and intra-operative findings. The median age at surgery was 5.2 years (range 1.6–15.8 years). Thirty-one patients underwent double patch repair, four patients a Warden repair, and two patients a single-patch repair. Of the four Warden repairs, two patients had a high insertion of right-sided anomalous pulmonary vein into the superior caval vein, one patient had bilateral superior caval veins, and one patient had right lower pulmonary vein insertion into the right atrium/superior caval vein junction. There was no post-operative mortality, reoperation, residual shunt or pulmonary venous obstruction. One patient developed superior caval vein obstruction and one patient developed atrial flutter. Conclusion: Complementary cardiac imaging modalities improve diagnosis of complex sinus venosus defects associated with a wide variation in the pattern of anomalous pulmonary venous connection. Nonetheless, surgical treatment is associated with excellent outcomes.

Introduction

Sinus venosus defect is a rare cause of interatrial communication representing up to 11% of all cases. Peacock described this defect permitting the interatrial communications as distinct from the normal atrial septum; hence, it is not a true atrial septal defect.[1] It has been well-established that the morphological criterion for diagnosis is the integrity of the rims of the oval fossa.[2,3] Previous authors described "unroofing" of the right pulmonary veins as the explanation for such defects, on the basis that a "shared wall" normally separated these venous structures from the superior caval vein and the cavity of right atrium.[4] However, this concept has been refuted as this "shared wall" is in fact an interatrial groove arising from an infolding of the atrial walls between the atrial connections of the caval and pulmonary veins, rather than a true atrial septum.[5,6] This aetiology was further validated by elegant episcopic microscopy studies.[7,8] In a review by Crystal et al. describing patients with inferior sinus venosus defects, the authors described the defining diagnostic feature as the anomalous connection of one or more pulmonary veins to the inferior caval vein, with the anomalous pulmonary vein or veins retaining their connection with the left atrium.[2] Recent anatomical studies have provided further evidence to support these defects representing veno-venous bridges allowing the interatrial communication.[7–10]

The pre-operative diagnosis of sinus venosus defects can be challenging and may be missed by solely relying on conventional planes in transthoracic echocardiography. This is due to the defects' close relationship with either the superior or inferior vena cava, outside the confines of the true interatrial septum.[11–13] Several studies have demonstrated echocardiographic subcostal sagittal-oblique bicaval view with use of colour flow mapping improves the detection rates of sinus venosus defects, notwithstanding the challenges in older obese patients due to poor echocardiographic windows.[2,3,14–18] Transoesophageal echocardiography is one of the imaging modalities of choice for this cardiac defect.[19–21] Other advanced cardiac imaging modalities such as cardiac MRI,[22] cardiac CT,[23,24] and rarely, cardiac catheterisation[11] have been used to

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complement the diagnosis of sinus venosus defects and delineation of anomalous pulmonary venous drainage.

The standard treatment for this defect has, up until recently, been surgical repair, although transcatheter closure in a selected group of patients is now increasingly an option.[25–32] Surgery aims to baffle the anomalously venous connection via the interatrial communication into the left atrium with or without enlarging the interatrial communication. Various surgical techniques have been well described with good results, including a single-patch baffle, double patch baffle, or Warden repair. However, there are associated low risks of sinus node dysfunction, atrial arrhythmias, and pulmonary or systemic venous occlusion after surgery.[33–44]

This study aimed to review the prevalence of complex sinus venosus defects defined as either the presence of multiple anomalous pulmonary venous sites or high insertion of the anomalous pulmonary vein into the superior caval vein, ascertain the imaging modalities employed in delineating the anatomical variants, and the surgical outcome in this cohort of patients.

Methods

All patients who had a diagnosis of sinus venosus defect with partial anomalous right pulmonary venous drainage who underwent surgical repair were identified retrospectively from a dedicated National Institute of Cardiology Outcomes Research database from 2008 to 2019. Each patient's medical records, diagnostic imaging modalities, and follow-up details were documented. Ethical approval was obtained from the Hospital Research Ethics Board.

Demographic and clinical details

Demographic details including patient age, gender, and weight at the time of the diagnosis and surgical repair were recorded. We documented the clinical symptoms at the initial presentation and the imaging modality used to reach the diagnosis. The primary outcome was procedural success (defined as successful completion of intended surgical procedure without revision). In addition, we noted any significant procedure-related complications.

Results

Patient demographic details are shown in Table 1. From 2008 to 2019, 37 children (20 male and 17 female) underwent surgical repair for sinus venosus defect with partial anomalous pulmonary venous drainage. Thirty-six of the patients have a diagnosis of superior sinus venosus defect and one patient was diagnosed with an inferior sinus venosus defect.

The median age at diagnosis was 4.2 years (range 0.5–15.5 years of age). In 32 of 37 (86%) patients, sinus venosus defect was diagnosed and partial anomalous pulmonary venous drainage were suspected or diagnosed by two-dimensional transthoracic echocardiogram including a subcostal sagittal-oblique bicaval view scanning from the sinus venosus defect to the anomalous pulmonary venous connection (Fig 1). In 5 of the 37 patients, the sinus venosus defect could not be visualised on transthoracic echocardiogram alone. In three of these five patients, who presented with unexplained right ventricular volume overload, the final diagnosis was achieved by transoesophageal echocardiogram (Fig 2); one of these three patients underwent further diagnostic cardiac catheterisation and cardiac CT. In two of these five patients, the diagnosis of sinus venosus defect and partial anomalous pulmonary venous drainage was reached intra-operatively. One of these two patients has an additional diagnosis of complex ventricular septal defect and the other was diagnosed with an inferior sinus venosus defect.

Variations of pulmonary venous drainage

The most common type of partial anomalous pulmonary venous drainage noted in this cohort was right upper and/or right middle pulmonary veins draining into the right superior caval vein. The patterns of insertion of the pulmonary veins are variable with low right superior caval vein being the commonest insertion point (19 of 37 patients). In ten cases, the anomalous pulmonary veins inserted into the right superior caval vein and right atrial junction (Fig 3). In three cases, the anomalous pulmonary veins drain directly into right atrium.

Five patients had complex pulmonary venous variants. Four patients had high insertion of anomalous vein into the superior caval vein (Fig 4) and three patients had multiple anomalous veins draining to different sites, two of whom had high drainage of one vein to the superior caval vein (Fig 5). Six of the 37 patients have bilateral superior caval veins.

Surgical management

The median age at the time of surgery was 5.2 years (range 1.6–15.8 years) and median weight was 21.7 kg (range 10.7–88 kg). Thirty-one patients underwent a double patch repair, four patients underwent a Warden repair, and two patients underwent a single-patch repair. Of the four patients who underwent a Warden repair, two of these four patients had a high insertion of right-sided anomalous pulmonary vein into the superior caval vein, one of these four patients had bilateral superior caval veins with relative hypoplastic right superior caval vein, and one patient had drainage of the right lower pulmonary vein to the right atrial and superior caval vein junction.

Follow-up

The median range of follow-up was 3.2 years (6 days–11.4 years). There was no mortality, reoperation, residual shunt, or pulmonary venous obstruction reported. One patient developed moderate degree of obstruction at the superior caval vein and right atrial junction at 8 months post-operatively; he subsequently underwent successful balloon dilatation to relieve the obstruction. One patient developed atrial flutter 2 months post-operatively and required cardioversion with restoration of sinus rhythm. No patient developed sinus node dysfunction.

Discussion

This study describes a heterogeneous group of children with sinus venosus defects, the majority of them were asymptomatic at presentation. The diagnostic sensitivity for sinus venosus defects was 86% (32 of 37) using transthoracic echocardiographic imaging including modified subcostal bicaval views. Transoesophageal echocardiography imaging was helpful in establishing the presence of sinus venosus defect in an additional three patients, increasing the total diagnostic yield of the echocardiographic examination from 86 to 94.5%. Transoesophageal echocardiographic imaging is proven to be a reliable supplementary diagnostic modality

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Table 1. Patient demographic details, patterns of anatomical variation, and surgical outcomes

Patient	Age at surgery (year)	Symptoms/signs	TTE	TOE	CMR	Others imaging modality	Operative findings	Type of repair	Complications
Group A: Superior SVD									
1	1.9	Murmur	SVD, Rt PAPVD	SVD, RUPV → R SCV	–	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
2	3.5	Intermittent tachypnoea	SVD, Rt PAPVD, bilateral SCVs	–	SVD, RU/RMPV → SCV/RA	–	SVD, RU/RMPV → SCV/RA. Smaller R SCV below the PVs	Warden repair	
3	2.8	Murmur	SVD, Rt PAPVD	–	SVD, RU/RMPV → R SCV	–	SVD, RU/RMPV → RA	Two-patch repair	
4	1.7	Intermittent cyanosis	SVD, normal PVs → LA	–	–	–	SVD, RUPV high insertion → R SCV	Warden repair	
5	4	FTT, effort intolerance,	SVD, suspected Rt PAPVD	SVD, Rt PAPVD	–	–	SVD, RUPV → R SCV	Two-patch repair	
6	12.5	DS, murmur, PDA	SVD, suspected Rt PAPVD	–	–	Diagnostic cath showed SVD, Rt PAPVD	SVD, RU/RMPV high insertion → R SCV at the level of azygous vein entry	Warden repair	
7	10.5	Effort intolerance	SVD, suspected Rt PAPVD	–	SVD, RU/RMPV→ SCV/RA	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
8	4.9	Murmur	SVD, suspected Rt PAPVD	SVD, All Rt PVs → R SCV	–	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
9	3.2	Murmur	SVD, Rt PAPVD	SVD, RLPV → RA	–	–	No intra-operative description	Warden repair	
10	1.7	Murmur	SVD, RUPV → RA	SVD, RUPV → RA	–	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
11	3.5	Murmur	SVD, Rt PAPVD	–	–	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
12	9.1	Murmur	SVD, Rt PAPVD, bilateral SCVs	–	SVD, RU/RMPV → SCV/RA	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
13	2.3	Intermittent cyanosis	SVD, Rt PAPVD	SVD, RUPV → SCV/RA	–	Diagnostic cath showed SVD, RUPV → SCV/RA	SVD, RU/RMPV →R SCV	Two-patch repair	
14	3.2	Murmur	SVD, Rt PAPVD, bilateral SCVs	–	SVD, RU/RMPV→ R SCV	–	SVD, RU/RMPV → R SCV	Two-patch repair	
15	15.8	rSR on ECG	SVD, suspected Rt PAPVD	SVD, RU/RMPV → R SCV	–	–	SVD, RU/RMPV → R SCV	Two-patch repair	
16	8	DCSA VSD	DCSA VSD	–	–	–	DCSA VSD, SVD, RU/RMPV → R SCV	Two-patch repair	
17	14.7	Murmur	SVD, suspected Rt PAPVD	SVD, Rt PAPVD	SVD, RU/RMPV → R SCV	–	SVD, RU/RMPV → R SCV	Two-patch repair	
18	10.3	Murmur	SVD, RUPV → RA	–	SVD, RUPV → R SCV	–	SVD, RU/RMPV → R SCV	Two-patch repair	

Table 1. (*Continued*)

19	6.2	2° ASD, BPA stenosis, RVVO	2°ASD, BPA stenosis, RVVO	2°ASD, SVD, RUPV → R SCV	–	–	2°ASD, SVD, RU/RMPV → R SCV	Two-patch repair	
20	13.8	Murmur, palpitation	SVD, suspected Rt PAPVD	–	SVD, RM/RLPV → R SCV	–	SVD, RU/RMPV → R SCV	Two-patch repair	
21	2.3	Murmur, LRTI	SVD, RU/RMPV →R SCV	SVD, RU/RMPV → R SCV	–	–	SVD, all Rt PVs → R SCV	Two-patch repair	
22	1.7	Murmur	SVD, suspected Rt PAPVD	SVD, All Rt PVs → RA	–	–	SVD, RU/RMPV → R SCV, RLPV → RA	Two-patch repair	
23	11.8	Effort intolerance	SVD, suspected Rt PAPVD	–	SVD, RUPV → R SCV	–	SVD, RUPV → R SCV	Two-patch repair	
24	1.2	CoA repair, progressive RVVO	SVD, suspected Rt PAPVD	–	–	Diagnostic cath showed SVD, All Rt PVs → SCV/RA	SVD, RUPV → R SCV	One-patch repair	
25	3.7	Murmur, LRTI	SVD, Rt PAPVD	SVD, Rt PAPVD	–	CTA showed SVD, RUPV → SCV/RA	SVD, All Rt PVs → R SCV	Two-patch repair	
26	6.5	2° ASD, BPA stenosis, RVVO	2° ASD, SVD, RUPV → SCV/RA	–	–	–	2° ASD, SVD, RUPV → SCV/RA	Two-patch repair	
27	15.6	Murmur	SVD, suspected Rt PAPVD	RU/RMPV → R SCV	–	–	SVD, RU/RMPV → R SCV	Two-patch repair	
28	11.7	Murmur	Mild PH, RVVO, bilateral SVCs	SVD, RU/RMPV → R SCV	–	Diagnostic cath showed SVD, RU/RMPV →SCV/RA; CTA showed SVD, RUPV → SCV/RA	SVD, RU/RMPV → R SCV	Two-patch repair	Developed atrial flutter 2 months post-op, reverted to sinus rhythm after synchronised DC cardioversion
29	8.7	Mild PS	Progressive RVVO	SVD, Rt PAPVD	–	–	SVD, RUPV → SCV/RA	Two-patch repair	
30	10.2	Murmur	SVD, Rt PAPVD, bilateral SCVs	–	SVD, RU/RMPV → SCV/RA	–	SVD, All Rt PVs → R SVC	Two-patch repair	
31	5.2	Murmur	SVD, Rt PAPVD	–	–	CTA showed SVD, RU/RMPV → R SCV	RU/RMPV → R SCV; 1 higher PV → R SCV	Two-patch repair	
32	4.8	Screening for FHx of CHD	SVD, Rt PAPVD, bilateral SCVs	SVD, RU/RMPV → R SCV	SVD, RU/RMPV → R SCV	–	SVD, RU/RMPV → R SCV	Two-patch repair	
33	7.4	Murmur, LRTI	SVD, RU/RMPV → R SCV	SVD, RU/RMPV → R SCV	SVD, RU/RMPV → R SCV	–	SVD, RU/RMPV → SCV/RA	Two-patch repair	
34	4.9	Cardiomegaly on CXR	SVD, suspected Rt PAPVD	–	–	CTA showed SVD, RUPV → SCV/RA	SVD, RU/RMPV → R SCV	Two-patch repair	SVC/RA obstruction which resolved post balloon dilatation
35	15.2	Screening for FHx of CHD	SVD, RUPV → SCV/RA	SVD, RUPV → SCV/RA	–	–	SVD, RU/RMPV → R SCV	Two-patch repair	

(*Continued*)

Table 1. (*Continued*)

Patient	Age at surgery (year)	Symptoms/signs	TTE	TOE	CMR	Others imaging modality	Operative findings	Type of repair	Complications
36	7.3	SVT, LRTI	SVD, suspected Rt PAPVD	SVD, RUPV → SCV/RA	SVD, RU/RMPV → R SCV	–	SVD, RU/RMPV → SCV/RA; 1 higher PV insertion → R SCV at the level of azygous vein entry	Two-patch repair	
Group B: Inferior SVD									
37	5.1	2° ASD	Progressive RVVO, 2°ASD	2°ASD with good anterior rim, absent posteroinferior or IVC rim; normal PVs → LA	–	–	Inferior SVD; RM/RLPV → RA	One-patch repair	

Abbreviations: →=to; 2°ASD=secundum atrial septal defect; BPA=branch pulmonary arteries; CHD=congenital heart disease; CMR=cardiac magnetic resonance imaging; CoA=coarctation; CTA=computer tomographic angiogram; CXR=chest X-ray; DCSA VSD=doubly committed subarterial ventricular septal defect; Diagnostic Cath=diagnostic cardiac catheterisation; DS=down syndrome; FHx=family history; FTT=failure to thrive; ICV=inferior caval vein; LA=left atrium; LRTI=lower respiratory tract infection; PDA=patent ductus arteriosus; PH=pulmonary hypertension; PS=pulmonary stenosis; R SCV=right superior caval vein; RA=right atrium; RLPV=right lower pulmonary vein; RM/RLPV=right middle and right lower pulmonary veins; RMPV=right middle pulmonary vein; Rt PAPVD=right partial anomalous pulmonary venous drainage; Rt PVs=right pulmonary veins; RU/RMPV=right upper and right middle pulmonary veins; RUPV=right upper pulmonary vein; RVVO=right ventricular volume overload; SCV/RA= right superior caval vein and right atrial junction; SCVs=superior caval veins; SVD=sinus venosus defect; SVT=supraventricular tachycardia; TOE=transoesophageal echocardiogram; TTE=transthoracic echocardiogram.

Figure 1. (movie clip): Transthoracic echocardiogram subcostal sagittal-oblique bicaval view scanning from the sinus venosus defect to the anomalous pulmonary venous connection.

particularly in the subgroup of patients who are obese or in the older patients, in whom a subcostal approach frequently provides images of a poor quality.[19,20]

This study highlighted a variation in preference for modality of the complementary imaging to aid confirmation of the diagnosis of sinus node defects, which is at the discretion of the attending paediatric cardiologist. With evolution in advanced cardiac imaging modalities, cardiac MRI and cardiac computer tomographic angiogram are increasingly used to aid diagnosis. Diagnostic cardiac catheterisation is rarely used nowadays to avoid radiation exposure.[11,22–24]

This study shows that the variation of the anomalous right pulmonary venous connections remains a diagnostic challenge. From this retrospective study and in the previous studies, [22–24] MRI (Fig 6) and CT imaging appear to be superior amongst imaging modalities in accurately delineating the anomalous pulmonary venous drainage, although the individual cardiologist's preference may vary between these modalities.

The surgical techniques used in our cohort of patients included a single-patch repair, double repair, and the Warden repair. The double patch repair was the dominant procedure performed. This is dependent on the complexity of the anatomy, anomalous pulmonary venous connections, and the surgeon's preference. A meta-analysis published by Okonta and Tamatey[45] looking at ten previous publications comparing the outcome of double- or single-patch repair reported that the adoption of double patch repair or the use of transcaval repair technique in single-patch repair was associated with a lower risk of venous obstruction. One patient (1/31 or 3%) from the double patch repair cohort from our study developed an obstruction at the superior caval vein and right atrial junction at 8 months post-operatively. He underwent successful balloon dilatation to relieve the obstruction subsequently. The incidence of superior caval vein obstruction in this study is comparable[46] or lower when compared to previous studies.[34,47] Post-operative sinus node dysfunction is more common in patients with single-patch or two-patch repair.[34,36,41,47,48] In this study, no patient developed sinus node dysfunction. Potential mechanisms causing sinus node dysfunction may include an anatomic anomaly of the sinus node and surgical trauma to the sinus node or to its blood supply.[48,49] Atrial arrhythmia is commonly reported both early and late after closure of these interatrial

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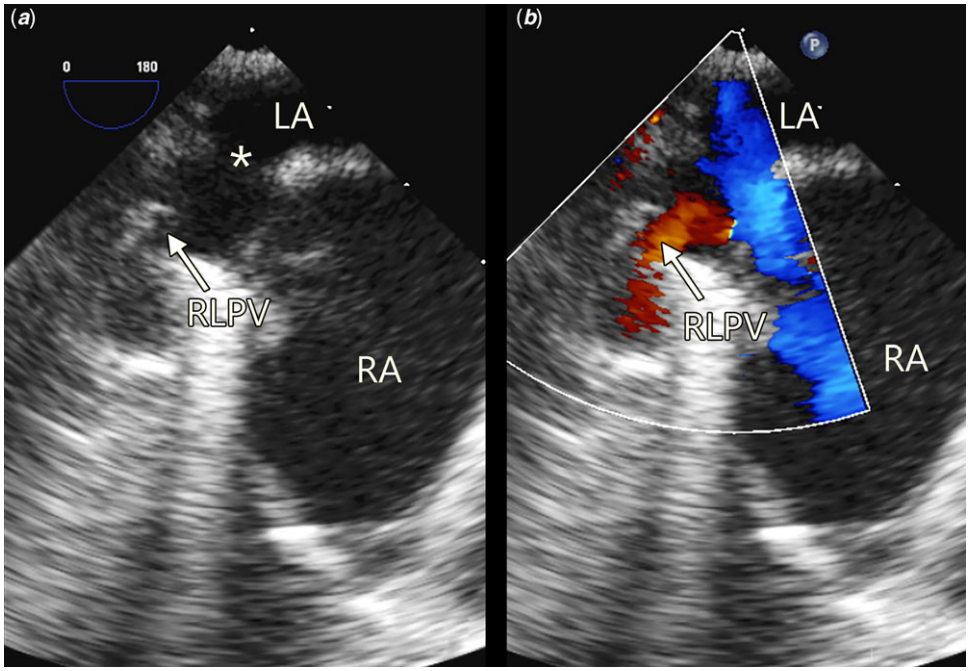

Figure 2. (***a*** and ***b***) Transoesophageal echocardiogram demonstrating the sinus venosus defect (*) and the anomalous right lower pulmonary vein (RLPV) draining directly into the right atrium (RA). LA=left atrium.

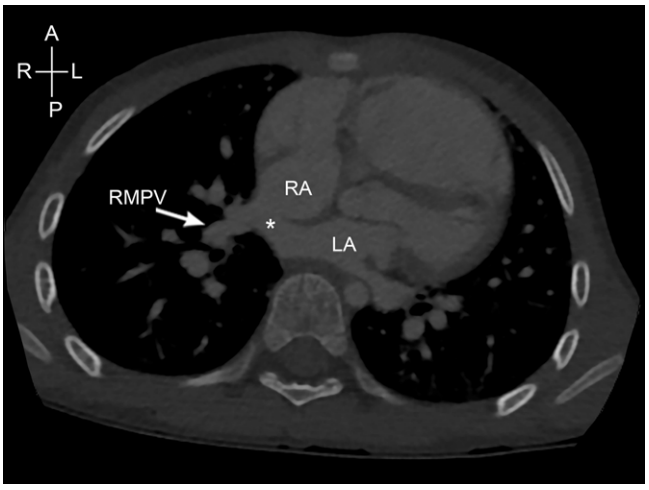

Figure 3. Computed tomography demonstrating right middle pulmonary vein (RMPV) draining to the right superior caval vein/right atrium junction. LA=left atrium; RA=right atrium.

communications.[34,47,48,50,51] One patient in this cohort developed late onset atrial flutter that responded to cardioversion. Potential mechanisms for atrial arrhythmia in these patients may include sinus node dysfunction with bradycardia-dependent atrial arrhythmias, scar-dependent multiple re-entries, and increased atrial size.

Although the surgical repair of this defect remains the gold standard, in recent years transcatheter closure has become an option in a select group of patients. In 2014, Garg et al.[26] reported a transcatheter closure of sinus venosus defect in a patient with bilateral superior caval veins, using a covered stent in the superior caval vein to close the defect and redirect the right upper pulmonary vein flow around the stent into the left atrium. With meticulous pre-procedural planning using balloon interrogation,[26–28] three-dimensional printed models,[25,29,30] image fusion guidance[31] or even holographic augmented reality,[32] these initial reports have demonstrated the feasibility and safety of transcatheter closure of sinus venosus defect in these patients in short to medium term (Fig 7). The transcatheter approach however may not be suitable in patients with right upper pulmonary vein that connects to high right superior caval vein or a relatively hypoplastic right superior caval vein especially in the presence of bilateral superior caval veins due to the risk of pulmonary venous obstruction by the covered stent.[29]

Conclusion

Complementary imaging modalities to transthoracic echocardiography are often required to delineate the exact pulmonary venous patterns in patients with sinus venosus defect. The pattern of anomalous pulmonary venous connection may be highly variable and complex in up to 14% of patients, including high insertion points and multiple drainage sites. Despite this, surgical treatment is associated with an excellent outcome and minimal complications. With progressive evolution in congenital cardiac catheterisation, a move to transcatheter closure has been shown to be suitable in select cases.

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Conflict of interest. None.

Ethical approval. This work complies with the ethical standards of the relevant national guidelines and with the Helsinki Declaration of 1975, as revised in 2008 and was approved by the Ethics committee at CHI Crumlin, Dublin, Ireland.

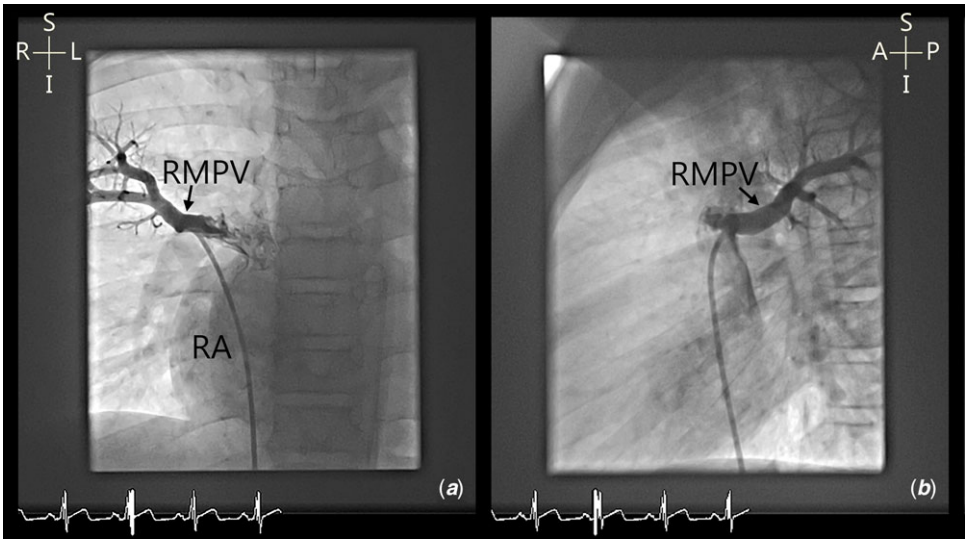

Figure 4. (***a*** and ***b***) Cardiac angiography demonstrating anomalous right middle pulmonary vein (RMPV) into right superior caval vein/right atrium junction. RA=right atrium.

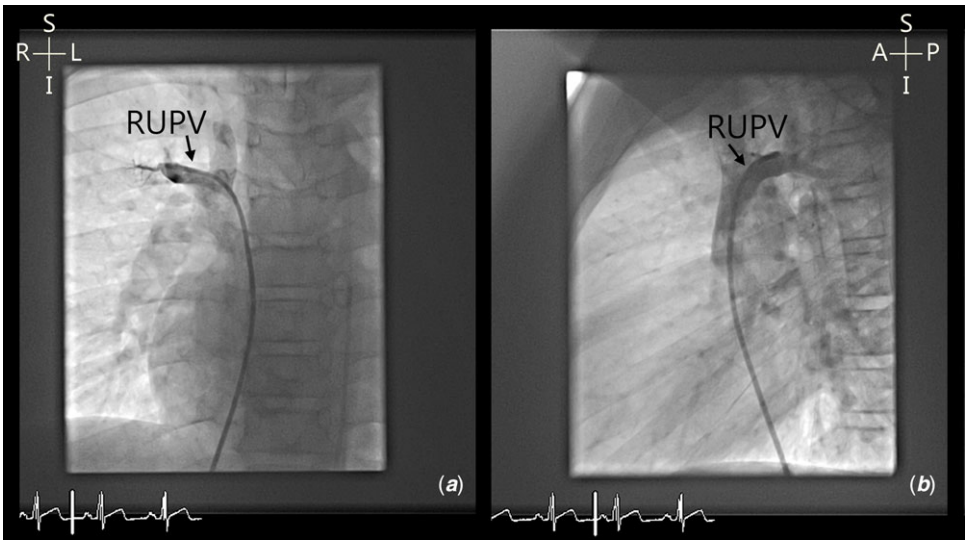

Figure 5. (***a*** and ***b***) Cardiac angiography demonstrating high insertion of right upper pulmonary vein (RUPV) into right superior caval vein.

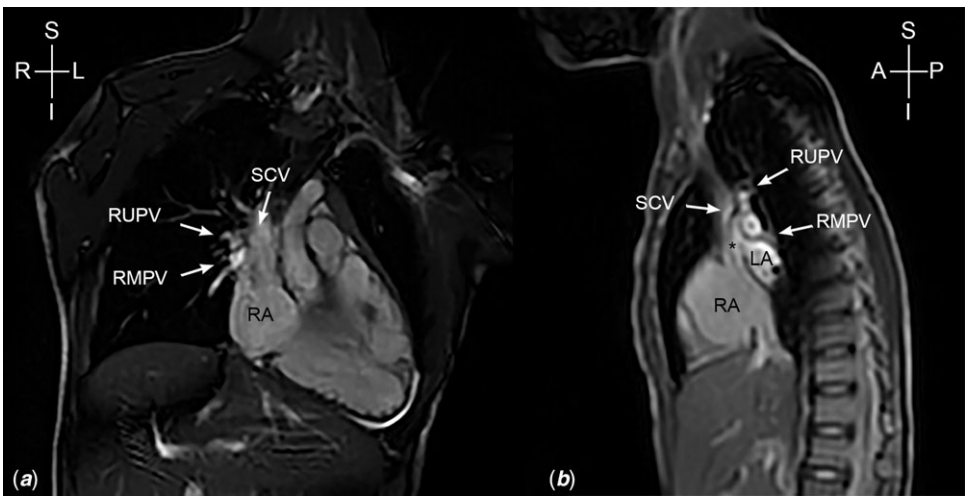

Figure 6. (***a*** and ***b***) Magnetic resonance angiography highlighting drainage of the right upper (RUPV) and right middle (RMPV) pulmonary veins draining into the right superior caval vein. RA=right atrium; LA=left atrium.

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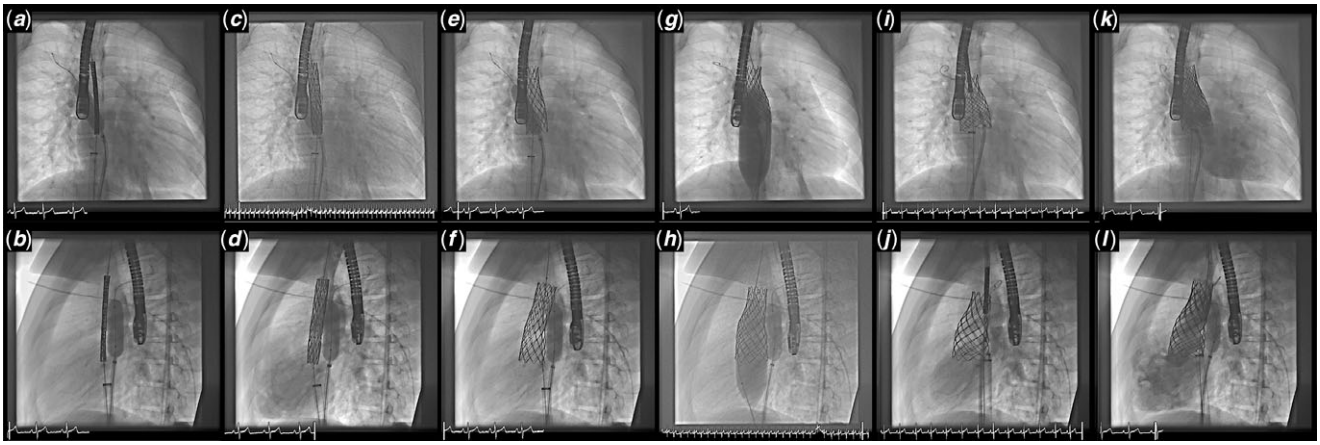

Figure 7. This series of angiography images show the steps in using a covered stent to repair a sinus venosus defect. (***a*** and ***b***) Anteroposterior (AP) and lateral views of a un-expanded 7 cm long 10-Zig covered CP stent mounted on a 28 mm BIB and introduced on a veno-venous guide wire rail and a separate pulmonary vein protection balloon (14 mm Atlas Gold balloon); (***c*** and ***d***) The inner balloon of a 28 mm 8 cm long BIB is expanded and the position adjusted on TOE imaging to overlap the crest of atrial septum; (***e*** and ***f***) The stent is further expanded by inflating the outer balloon; (***g*** and ***h***) Further flaring of the lower half of the stent is performed until the stent abuts the septum and there is no or minimal residual shunt on TOE. (h and i) An un-expanded bare-metal anchor stent and (k and l) post-expanded bare-metal anchor stent is the place to secure the flared conical Covered CP stent. The pulmonary vein is protected throughout by keeping the high pressure balloon inflated inside the pulmonary vein whenever the covered stent is dilated.

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