Cardiology in the Young

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Leftward deviation of the primary septum or dividing left atrial shelf?

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Review

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Abstract

Isolated leftward prolapse or deviation of the primary atrial septum is a rare CHD that can mimic abnormal pulmonary venous return at first sight. We present a case of a newborn infant, referred for surgical correction of totally anomalous pulmonary venous return into the right atrium, with the peri-operative finding of a leftward deviation of the superior margin of the primary atrial septum. The distinction with a dividing atrial shelf could not be confirmed with certainty. Fifty-three similar cases from the literature are incorporated. A detailed review of the current account on atrial septation is studied. The embryological and clinical features of a dividing partition of the left atrium are discussed.

Leftward prolapse or deviation of the primary atrial septum is a CHD characterised by displacement of the superior margin of the primary atrial septum well into the left atrium, the base of the primary septum being normally formed. This results in the pulmonary venous flow to the right atrium, despite the anatomic connection of the pulmonary veins to the left atrial wall.[1] It thereby, depending on the degree of deviation, mimics a partial or totally anomalous pulmonary venous return. Additionally, the flap valve of the oval fossa divides the left atrium, much like as in a divided morphologically left atrium[2] and a "fenestration" or a paraseptal leak is necessary to allow blood flow into the left ventricle. The differential diagnosis between these different anatomical conditions may be difficult. We present a case of a newborn infant referred for surgical correction of totally anomalous pulmonary venous return, but with the peri-operative finding of an isolated leftward prolapse of the primary septum, or was it a dividing left atrial shelf? Subsequently, embryological and anatomical features of the atrial septum are discussed. Clinical features from another 53 cases from the literature are presented.

Case report

A 6-week-old infant presented to the paediatric team with feeding difficulties and was found on examination to have discrete tachypnoea, a systolic ejection murmur on auscultation and transcutaneous oxygen saturation of 92% in room air. Initial transthoracic cardiac echography suspected totally anomalous pulmonary venous return. The baby was referred for cardiac surgery. Biometry and ECG were normal. Repeat cardiac echography (GE Healthcare Vivid E90 Ultrasound System; Fig 1) showed, in the context of situs solitus, disproportion between the left and right heart structures with, although reaching the apex, a small left ventricle. The M-mode-based left ventricle end-diastolic diameter measured 14.8 mm, corresponding to −3.7 Z-score (Heart Center Z-score calculator, Boston Children's Hospital) and the right ventricle end-diastolic diameter 23.6 mm. The 4-chamber measurement of the mitral valve size was 9 mm (−2.1 Z-score) with no signs of dysfunction; however, the tricuspid valve measured 16.5 mm (+2.4 Z-score) with mild regurgitation. An enlarged pulmonary valve (annulus diameter of 15 mm, +2.4 Z-score) with turbulent high flow was observed, in contrast to a 6.4 mm aortic valve annulus diameter (−1.9 Z-score). The transverse aortic arch measured 7 mm (−0.37 Z-score) and the aortic isthmus diameter 4 mm (−2.0 Z-score). The pulmonary arteries measured 7 mm (+1.0 Z-score) and 6 mm (+0.2 Z-score), right and left, respectively.

Above both ventricles, a large atrial cavity, measuring 25 × 23 mm, was visualised, receiving systemic veins from the right side and pulmonary veins from the left side. Using colour Doppler, significant flow from the pulmonary veins was visualised, entering the cavity and reaching the right side of the cavity. Furthermore, a connection between the large atrial and a minute supra-mitral cavity, delineated by a 4 mm fenestrated membrane, was seen on colour Doppler. Restrictive and turbulent flow was evident between the large and smaller cavities. At first sight, the large atrial cavity was interpreted as an enlarged right atrium and the small supra-mitral cavity as left atrium, with pulmonary flow above the left atrium towards the right atrium and thus totally anomalous pulmonary venous return.

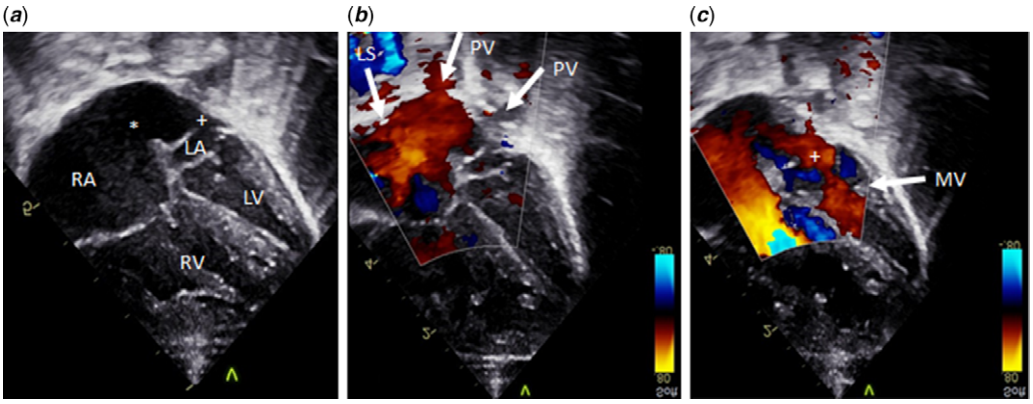

Figure 1. Pre-operative transthoracic echocardiographic findings in our patient, subcostal 4-chamber views. (***a***) Bidimensional subcostal 4-chamber view showing significant leftward orientation of the primary septum and small left heart structures with a dilated RA and RV; (***b***) colour subcostal 4-chamber view showing a hypoplastic limbus superior and normal pulmonary venous connection to the left side of the atrial body; (***c***) colour subcostal 4-chamber view showing an atrial communication between the primary septum and the posterior wall of the LA permitting anterograde blood flow through the mitral valve. * = atrial septum defect; + = fenestrated displaced flap valve of the oval fossa; LA = left atrium; LS = limbus superior; LV = left ventricle; MV = mitral valve; PV = pulmonary vein; PV = pulmonary veins; RA = right atrium; RV = right ventricle.

On closer inspection, however, the superior interatrial fold seemed hypoplastic with a very small rim superiorly in the middle of the larger atrial cavity. The membrane between the atrial cavities represented to us the primary septum, commonly called the flap valve of the oval fossa, shifted leftwards and attached on the posterior left atrial wall, creating an image of divided left atrium. No additional imaging was performed.

During surgical inspection, there was the impression of a mono-atrium, with drainage of all four pulmonary veins into the left wall of the large atrial cavity. The superior part of the primary septum was indeed visualised as a membrane right above the mitral valve, dividing the left atrium as in a divided morphologically left atrium or so-called "cor triatriatum sinister." This partition divided the left atrium sub-totally with minimal shunt laterally and through a fenestration. After inspection, the partition was detached and mobilised towards its normal position and fixated by a simple suture to the small infolded superior rim in the atrial roof. Fenestration was closed by a single suture. Given the normal anatomical drainage of all four pulmonary veins, no rerouting was performed. The post-operative course was uncomplicated. Cardiac ultrasound confirmed complete closure of the interatrial septum, normal pulmonary venous return into the left atrium and unobstructed left ventricle inflow. Further follow-up was uneventful.

Discussion

Clinically significant leftward prolapse of the primary septum was described by Van Praagh et al.[1] Usually, it is associated with hypoplastic left heart syndrome or in isomerism syndromes.[1,3–5] Few cases of isolated forms in young infants such as our patient have been described. Understanding the embryology of the normal atrial septum development as well as reviewing the case studies existing of this malformation assist in the understanding of this anatomic abnormality.

Embryological features of atrial septation

Firstly, in reviewing the morphogenenis of leftward prolapse of the primary septum, an appropriate account of the embryology of atrial septation should be discussed. Atrial septal development is a complicated topic and largely described in the literature. Based on Anderson's work,[6] the theory that the secondary atrial septum arises from the atrial roof and overlaps the primary septum is inaccurate. Indeed, the septation of the atrial component of the heart initiates from a ridge of mesenchyme in the roof of the undivided atrial cavity. We first see the appearance of the primary atrial septum, which grows as an interatrial shelf. At that time, endocardial cushions have developed within the atrioventricular canal. These cushions grow towards each other to divide the canal, whilst the primary septum grows towards the cushions. By this movement, the cranial portion of the septum, at its origin from the atrial roof, has broken down, creating the secondary interatrial foramen. This hole is an essential part of the foetal circulation, permitting the placental oxygenated rich blood to return and reach the left side of the developing systemic circulation. Once the pulmonary veins are incorporated into the atrial roof, the upper margin of this foramen become converted into the interatrial fold. The superior rim of the oval fossa, better described as the superior interatrial fold, is thus a false septum, although commonly described as the secondary atrial septum.

A potential ambiguity in the terminology currently employed by Anderson exists at this stage. The superior fold can be interpreted as representing a "buttress" against which the flap valve will abut so as to close the oval fossa. Nonetheless, the current account of septal development endorsed by Jensen et al[7] emphasises that there is a true second atrial septum involved in the atrial septation. The second atrial septum originates from the vestibular spine and then muscularises antero-inferiorly to form the buttress for the flap valve of the definitive oval fossa, the flap itself being formed by the primary atrial septum. This version leaves the superior interatrial fold as a superior buttress and the second septum as an antero-inferior one. After birth, the pulmonary venous drainage into the left atrium dramatically increases, and the right aspect of the primary septum, represented by the flap valve of the oval fossa, is pushed against (and must fuse with) the left aspect of the superior interatrial fold, closing the oval fossa. The true second atrial septum anchors the flap valve to the atrioventricular junctions.

Van Praagh's description of the development of the atrial septum is different.[1,2] The infolding of the atrial roof results in a superior limbic band, being part of the secondary atrial septum. It is against this limbic band that the cephalic border of the primary

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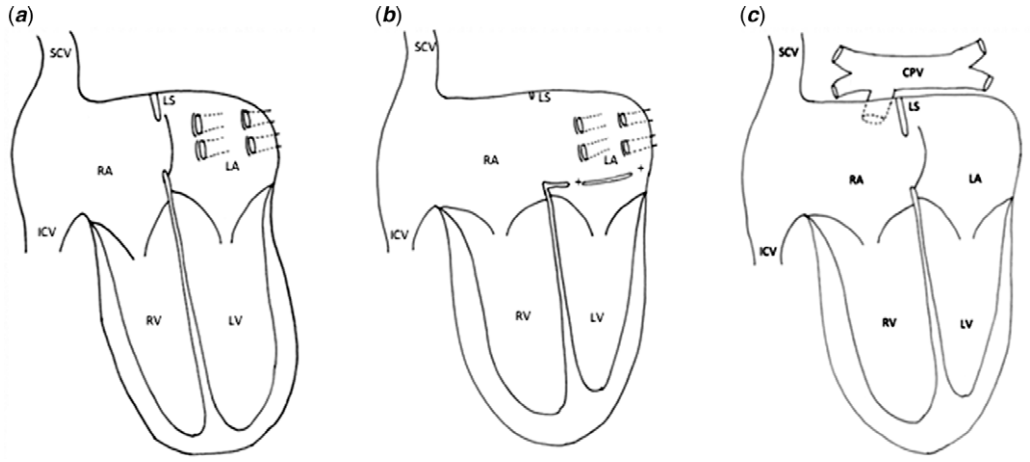

Figure 2. Schematic presentation of cardiac anatomy in differential diagnosis, ventricular outflow tracts and great vessels not represented. (***a***) Normal cardiac anatomy at birth; (***b***) leftward prolapse of the primary septum, (***c***) intracardiac totally anomalous pulmonary vein return. + = fenestrated displaced flap valve of the oval fossa; CPV = common pulmonary vein; ICV = inferior caval vein; LA = left atrium; LS = limbus superior; LV = left ventricle; RA = right atrium; RV = right ventricle; SCV = superior caval vein.

septum normally attaches. However, to our knowledge, there is no evidence from developing hearts that a second septum grows from the atrial roof. Anyway, normal development of the superior folding is essential for the atrial septation. If the superior limbic band is hypoplastic, resulting from an insufficient infolding of the atrial roof, it provides insufficient support to the superior border of the primary septum allowing prenatal prolapse towards the left atrium driven by the foetal blood flow.[1] If the flap valve of the primary septum deviates beyond the pulmonary veins, postnatal increase of the pulmonary venous return will not push the primary septum against the superior interatrial fold and will not close the oval fossa (Fig 2a and b).

Clinical features

Secondly, we conducted a systematic search to identify all cases published with deviation of the primary septum causing abnormal pulmonary venous flow. The following search terms were used to search for relevant articles in PubMed: ("Atrial Septum"[Mesh] OR atrial-septum [tiab] OR septum-primum [tiab]) AND (Malposition [tiab] OR displacement [tiab] OR attachment [tiab] OR deviation [tiab] OR prolapse* [tiab]). All English articles describing one or more cases of deviation of the primary septum causing abnormal pulmonary venous return were suitable for inclusion. Articles were only included if the full text was available online. The following data were extracted from the articles: author(s), year of publication, age of the patient(s), presenting symptoms, situs, aspect of the superior, partial or total abnormal pulmonary venous drainage, associated cardiac pathology, treatment and outcome. In total, 126 articles were identified. Based on title and abstract, 110 articles were excluded. The full text of 16 articles was available for review and 8 articles were suitable for inclusion. In total, 53 cases were described in detail. An overview of these cases is given in Table 1. From this analysis, we concluded that age at diagnosis varied between 0,3 and 120 months. Potential symptoms at presentation varied with showing signs of cardiac failure and other being asymptomatic. Common features mentioned are transcutaneous desaturation, dyspnoea, arrhythmia and heart murmurs.

The diagnosis can be confirmed using two-dimensional transthoracic cardiac echography, with following features being key signs for the diagnosis: leftward displacement of the primary septum, dividing the left atrium, thereby allowing half or all of the pulmonary veins to drain into the right atrium, despite their anatomic connection to the left atrial wall; hypoplastic or absent infolded superior rim; small appearance of the left ventricle and left atrium beneath the flap; enlargement of the right atrium and ventricle; secondary interatrial communication.

Van Praagh called it a "primary septum malposition defect" as it presents an interatrial communication associated with the (partial) absence of the secondary atrial septum and malposition of primary septum.[1,2] In fact, in divided left atrium, the interatrial communication can be open either to the pulmonary or the vestibular component of the divided atrium. In either event, however, it is the secondary interatrial communication. This is because the communication is between the cranial edge of the primary atrial septum, or the flap valve, and the roof of the atrial chambers. The position of the communication does not change its developmental heritage. It is the anatomical borders that determine the phenotypic specificity. One or more fenestrations in the primary septum can coexist, as in our case.

It is clear from an echocardiographic point of view, leftward prolapse of the primary septum may be misdiagnosed.[2,17] The images can be similar to intracardiac partial or totally anomalous pulmonary venous return into the right atrium. In our case, because all four pulmonary veins drained into the left-sided atrial wall, this condition could not be considered as totally anomalous pulmonary venous return, even if the drainage is above the primary septum and in terms of physiology, similar to totally anomalous pulmonary venous return (Fig 2c). Moreover, it is also exceedingly rare to find totally anomalous pulmonary venous connection directly to the morphologically right atrium other than in the setting of right isomerism.

Regarding the entity of divided left atrium, it is defined by an obliquely orientated fibromuscular partition dividing the morphologically left atrium.[9] There are many classifications described with high variability in the position of the pulmonary veins and the position of the communication between the compartments. The morphogenesis of the divided left atrium is conventionally explained on the basis of failure of absorption of the common pulmonary vein into the left atrium. Another hypothesis is the "entrapment concept" of the primary pulmonary vein, in combination with "malseptation," if the atrial septum defect

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Table 1. Published clinical cases with deviation of the septum primum causing abnormal pulmonary venous return (or drainage).

Case	Article	Age (months)	Symptoms	Situs	Limbus superior	Pulmonary venous return	Other pathology	Diagnosis	Treatment	Outcome
1	Hubrechts et al	1.5	Dyspnoea, heart murmur	Solitus	Hypoplastic	TAPVD	None	Peroperative	Atrial septal displacement	Discharged
2	Ayyildiz et al[8]	24	–	Solitus	Absent	PAPVD (RUPV)	VSD	TTE	Pericardial patch	Discharged
3	Ayyildiz et al[8]	12	–	LAI	Absent	TAPVD	LPSVC ASD	TTE	Atrial septal displacement	Discharged
4	Ayyildiz et al[8]	120	–	Solitus	Absent	PAPVD (RUPV)	Cor triatriatum ASD	TTE	Pericardial patch	Discharged
5	Ayyildiz et al[8]	24	–	Solitus	Absent	PAPVD (RUPV)	None	TTE	Not operated	Discharged
6	Ayyildiz et al[8]	60	–	Solitus	Absent	PAPVD (RUPV + RLPV)	ASD	TTE	Atrial septal displacement	Discharged
7	Ayyildiz et al[8]	12	–	LAI	Absent	PAPVD (RUPV)	LPSVC ASD	TTE	Pericardial patch	Discharged
8	Ayyildiz et al[8]	18	–	LAI	Absent	PAPVD (RUPV + RLPV)	ASD	TTE	Atrial septal displacement	Discharged
9	Ayyildiz et al[8]	42	–	Solitus	Absent	PAPVD (RUPV)	None ASD	TTE	Pericardial patch	Discharged
10	Ayyildiz et al[8]	6	–	Solitus	Absent	PAPVD (RUPV + RLPV)	VSD ASD Cor triatriatum	TTE	Pericardial patch	Discharged
11	Ayyildiz et al[8]	31	–	Solitus	Absent	PAPVD (RUPV)	ASD	TTE	Pericardial patch	Discharged
12	Ayyildiz et al[8]	27	–	Solitus	Absent	PAPVD (RUPV + RLPV)	ASD	TTE	Pericardial patch	Discharged
13	Ayyildiz et al[8]	5	–	LAI	Absent	TAPVD	VSD ASD LV hypoplasia	TTE	Pericardial patch	Discharged
14	Ayyildiz et al[8]	2	–	LAI	Absent	PAPVD (RUPV + RLPV)	LV hypoplasia	TTE	Atrial septal displacement	Discharged
15	Ayyildiz et al[8]	12	–	LAI	Absent	TAPVD	ASD DORV LPVSC	TTE	Pericardial patch	Died
16	Ayyildiz et al[8]	12	–	Solitus	Absent	PAPVD (RUPV + RLPV)	AVSD	TTE	Pericardial patch	Discharged
17	Jhaveri et al[13]	60	Heart murmur	Inversus totalis	Absent	PAPVD (LUPV + LLPV)	BSVCV Dextrocardia	TTE	Pericardial patch	Discharged

Table 1. (*Continued*)

18	Cuttone et al[2]	66	Severe dyspnoea	Solitus	Absent	TAPVD	ASD	TTE	Pericardial patch	Discharged
19	Cuttone et al[2]	78	Dyspnoea	Solitus	Absent	TAPVD	ASD	TTE	Pericardial patch	Discharged
20	Cuttone et al[2]	3	Cardiac failure Sinus bradycardia	Solitus	Absent	TAPVD	ASD	TTE	Pericardial patch	Discharged
21	Prasad et al[14]	24	Dyspnoea	Solitus	Absent	PAPVD (RUPV + RLPV)	Inferior sinus venosus	TTE	Atrial septal displacement	Discharged
22	Atik et al[15]	11	Dyspnoea	Solitus	–	TAPVD	–	Peroperative	Patch	Discharged
23	Tomar et al[16]	3	–	LAI	Absent	PAPVD (RUPV + RLPV)	IVC interruption* BSVCV	TTE	–	–
24	Tomar et al[16]	1	–	LAI	Absent	TAPVD	Dextrocardia IVC interruption* DORV PS	TTE	–	–
25	Tomar et al[16]	16	–	LAI	Absent	PAPVD (RUPVD + RLPVD)	IVC interruption*	TTE	Surgery (not otherwise specified)	–
26	Tomar et al[16]	4	–	Solitus	Absent	TAPVD	DORV VSD PS	TTE	–	–
27	Tomar et al[16]	6	–	Solitus	Absent	TAPVD	None	TTE	Surgery (not otherwise specified)	–
28	Tomar et al[16]	15	–	RAI	Absent	TAPVD	Common AV valve VSD PS BSVCV	TTE	–	–
29	Tomar et al[16]	36	–	Solitus	Absent	TAPVD	None	TTE	–	–
30	Tomar et al[16]	0,3	–	Solitus	Absent	TAPVD	None	TTE	–	–
31	Tomar et al[16]	0,6	–	LAI	Absent	TAPVD	Dextrocardia IVC interruption* DORV PS	TTE	–	–
32	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	ASD	TTE	DASP removal ASD closure	Alive and well
33	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	ASD	TTE	DASP removal ASD closure	Alive and well

(*Continued*)

Table 1. (*Continued*)

Case	Article	Age (months)	Symptoms	Situs	Limbus superior	Pulmonary venous return	Other pathology	Diagnosis	Treatment	Outcome
34	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	ASD Mitral valve prolapse	TTE	None	Alive and well
35	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Heterotaxia Atrioventricular canal Subaortic stenosis Coarctation aorta Complete heart block	TTE	None	Died awaiting heart transplant
36	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Heterotaxia Tetralogy of Fallot	TTE	Tetralogy repair	Died after repair
37	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Heterotaxia Atrioventricular canal Subaortic stenosis	TTE	Glenn anastomosis DASP removal	Died after Glenn
38	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Heterotaxia ASD	TTE	ASD repair Patch PV to LA	Alive and well
39	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Heterotaxia Transposition of the great arteries Atrioventricular canal	TTE	DASP removal Fontan at 11 years	Alive and well
40	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	VSD Polyvalvular disease	TTE	None	Died
41	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	VSD Arch hypoplasia	TTE	VSD closure Arch augmentation	Lost to follow-up
42	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	VSD	TTE	None	Alive and well
43	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	VSD	TTE	None	Lost to follow-up
44	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	VSD	TTE	None	Alive and well
45	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Coarctatio aortae	TTE	Coarctation repair	Alive and well
46	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Critical aorta stenosis	TTE	Aortic valve dilation, converted to Norwood DASP removal	Died after Fontan operation
47	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Tetralogy of Fallot with pulmonary atresia	TTE	None	Lost to follow-up
48	Cohen et al[17]	–	–	–	–	PAPVD (RUPVD + RLPVD)	Transposition of the great arteries Tricuspid atresia	TTE	Fontan operation DASP removal	Lost to follow-up

Table 1. (*Continued*)

49	Hiramatsu et al[11]	1	–	–	Absent or hypoplastic	TAPVD	VSD ASD	TTE or catheterisation +	Atrial septal displacement	–
50	Hiramatsu et al[11]	12	–	–	Absent or hypoplastic	PAPVD	Polysplenia PS ASD	TTE or catheterisation+	Atrial septal displacement	–
51	Hiramatsu et al[11]	4	–	–	Absent or hypoplastic	TAPVD	PDA ASD	TTE or catheterisation+	Atrial septal displacement	–
52	Hiramatsu et al[11]	7	–	–	Absent or hypoplastic	PAPVD	ASD	TTE or catheterisation+	Atrial septal displacement	–
53	Hiramatsu et al[11]	36	–	–	Absent or hypoplastic	PAPVD	ASD	TTE or catheterisation+	Atrial septal displacement	–

ASD = atrial septal defect; AVSD = atrioventricular septal defect; BSVCV = bilateral superior vena caval veins; DASP = deviation of the septum primum; DORV = double outlet right ventricle; IVC = inferior vena cava; LAI = left atrial isomerism; LLPV = left lower pulmonary vein; LPSVC = left persistent superior caval vein; LUPV = left upper pulmonary vein; LV = left ventricle; PAPVD = partial pulmonary venous drainage; PDA = patent ductus arteriosus; RAI = right atrial isomerism; RLPV = right lower pulmonary vein; RUPV = right upper pulmonary vein; TAPVD = total abnormal pulmonary venous drainage; TTE = trans thoracic echocardiography; VSD = ventricular septal defect.
*with azygos continuation.
+Three out of five cases were diagnosed using TTE. Two out of five cases were diagnosed using catherisation.

communicates with the proximal chamber.[9] Cuttone et al believe that the "primum movens" of leftward prolapse of the primary septum is different, with failure of the development of the superior limbic band of the secondary atrial septum, insisting on a different position of the atrial septal defect, in a low position near the atrioventricular junction in the classical form of divided left atrium. In leftward displacement of the primary septum, the communication is higher, far from the atrioventricular junction.[2] These theories, on differentiating divided left atrium from leftward prolapse of the primary septum, are speculative. None of the previous investigators have provided any evidence to prove the true nature of the partition. In our clinical case as in most of the reviewed literature cases (Table 1), there is no histologically evidence neither to confirm that the partition dividing the cavity of the left atrium is the deviated primary atrial septum, and not a dividing left atrial shelf as in the setting of a divided morphologically left atrium. In future cases, if it proved possible to remove the partition and study it histologically,[10] it should be possible to determine whether it is the primary septum or a dividing shelf.

Correct diagnosis however facilitates surgical management. When available and feasible, pre-operative 3D-transthoracic echocardiography is a valuable tool to visualise the intracardiac anatomy and to determine the operative strategy.[2] The most frequently used surgical technique to correct a leftward prolapse of the primary septum consists of a complete resection of the malpositioned flap valve followed by the reconstruction of an appropriately positioned septum using either pericardium or prosthetic material.[1] It is important to perform a complete resection of the primary septum to prevent pulmonary venous obstruction.[11,12] Another technique, namely performing a septoplasty using the original primary septum, has been described to avoid patch retraction and to allow natural septal growth.[1,11] Otherwise, as re-siting the primary septum, repositioning of a dividing shelf is also feasible. This second technique was used with success in our case. This technique is however often not applicable as the partition (dividing shelf or primary septum) may be severely displaced and malformed or adherent to the left atrial wall.[2] Prognosis of isolated forms of leftward prolapse of the primary septum is expected to be very good as no pulmonary venous rerouting is necessary.

Conclusion

Leftward prolapse of the primary septum is considered as the deviation of the superior margin of the primary atrial septum, dividing the left atrium. Pulmonary veins are externally normally connected to the left atrial wall, but the drainage of pulmonary venous flow visually goes into the right atrium. This may be incorrectly diagnosed as partial or totally anomalous pulmonary venous return. Moreover, there is currently no evidence to substantiate that the described partition is initially a deviated primary septum rather than a dividing atrial shelf. The existing theories for the development of the atrial septum are not able to answer the question about the true nature of the partition in this type of divided left atrium. Correct diagnosis, however, is important to facilitate surgical repair and to predict prognosis.

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