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# Endocardial transvenous pacing in patients with surgically palliated univentricular hearts: A review on different techniques, problems and management

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## ABSTRACT

Fontan surgery and its modifications have improved survival in various forms of univentricular hearts. A regular atrial rhythm with atrioventricular synchrony is one of the most important prerequisite for the long-term effective functioning of this preload dependent circulation. A significant proportion of these survivors need various forms of pacing for bradyarrhythmias, often due to sinus nodal dysfunction and sometimes due to atrioventricular nodal block. The diversion of the venous flows away from the cardiac chambers following this surgery takes away the simpler endocardial pacing options through the superior vena cava. The added risks of thromboembolism associated with endocardial leads in systemic ventricles have made epicardial pacing as the procedure of choice. However challenges in epicardial pacing include surgical adhesions, increased pacing thresholds leading to early battery depletion and frequent lead fractures. When epicardial pacing fails, endocardial lead placement is equally challenging due to lack of access to the cardiac chambers in Fontan circulation. This review discusses the univentricular heart morphologies that may warrant pacing, issues about epicardial pacing, different techniques for endocardial pacing in patients with disconnected superior vena cava, pacing in different modifications of Fontan surgeries, issues of systemic thromboembolism with endocardial leads, atrioventricular valve regurgitation attributed to pacing leads and device infections. In a vast majority of patients following Glenn shunt and Senning surgery, an epicardial pacing and lead replacement is always feasible though technically very difficult. This article highlights the different options of transatrial and transventricular endocardial pacing.

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## 1. Introduction

### 1.1. Bradyarrhythmia in congenital heart diseases

Various congenital heart diseases and their surgical treatments are associated with bradyarrhythmias. Heterotaxy syndromes, atrioventricular discordance, single ventricles are more often associated with these rhythm disorders than others. Pacemaker implantation is the commonest reintervention following Fontan surgery due to sinus node dysfunction in 9–23% of patients and atrioventricular nodal conduction abnormalities in 2–16% of patients [1–3]. Atrioventricular nodal block may occur either

spontaneously in the natural history of few univentricular hearts or follow treatment of atrial tachyarrhythmia. It may sometimes follow certain surgical procedures like enlargement of bulboventricular foramen during Fontan surgery [4]. Technological advances in pacemaker designs, improving operator experience and miniaturization of pulse generators and leads have increased the number of young patients who get permanent pacing for bradyarrhythmias [5]. However the small size of pediatric patients and frequent venous occlusions after pacing discourage against very early institution of pacing therapy.

### 1.2. Pacing in Fontan circulation

Fontan surgery and its modifications, employed in different univentricular hearts involve diversion of the caval flows towards the pulmonary artery. The superior vena caval diversion following Fontan surgery has made epicardial pacing as the preferred strategy

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in these patients [6]. Its limitations include high pacing thresholds leading to short battery life, frequent lead fractures in 17–40% at five years after implantation [7]. Surgical adhesions increase the technical difficulties to implant the leads in the epicardium. Even though endocardial leads are superior to epicardial leads in their performance, lack of access to heart for placing these leads and increased risk of systemic thromboembolism pose challenges to endocardial pacing [8].

### 1.3. Thrombus formation over the endocardial leads

The risk of systemic thromboembolism due to endocardial leads in the systemic ventricles discouraged endocardial pacing in univentricular hearts [9]. Fontan surgery adds to this risk due to sluggish blood flows in the absence of a right ventricular pump, increased venous pressure causing increased factor VIII levels, the presence of prosthetic material, and potential protein C deficiency [10]. Incidence of thromboembolism in the Fontan circulation varies from 3 to 20%. Routine anticoagulation reduces the risk by more than 50% and is recommended for life in many institutions following Fontan surgery despite controversies [10,11]. Patients with intracardiac shunts have a two-fold increase in risk of systemic thromboembolism when they have transvenous endocardial pacing or defibrillation leads [12]. It is mandatory to strongly consider lifelong adequate anticoagulation in every patient who has an endocardial lead placed in a systemic ventricle in Fontan circulation.

### 1.4. Endocardial access following Fontan surgery

The most important reason that precludes endocardial pacing in patients following Fontan surgery is the lack of an easy access to the cardiac chambers following the venous diversion caused by this surgery [13]. A Glenn anastomosis connects the superior vena cava directly to the pulmonary artery and removes the possible access from subclavian veins to the different cardiac chambers. This review details a few clinical scenario that warranted innovative techniques (Fig. 1) to achieve an endocardial access for pacing. A brief literature review on these techniques also follows in the discussions.

## 2. Case scenario

### 2.1. Transhepatic pacing

A seven-year-old male with left isomerism, azygos continuation of interrupted inferior vena cava, common atrioventricular valve and univentricular heart underwent Total cavopulmonary shunt (Kawashima repair). His preoperative electrocardiogram showed complete heart block with ventricular rate of 45/minute. Epicardial pacing failed in the post-operative period with very high pacing thresholds in spite of placing the temporary pacing lead in different epicardial locations. A *trans*-hepatic endocardial lead was placed in the ventricle and connected to a permanent generator in the sub-rectus sheath in the anterior abdominal wall (Fig. 2). A midline liver that is a characteristic of heterotaxy syndromes permitted ultrasound guided puncture into the hepatic vein through subxiphoid access. After placing a sheath over a guidewire into the atrium, a screw-in endocardial lead was manipulated into the ventricle and securely positioned with a loop in the atrium to allow for growth of the child. A snug-fitting 6F sheath was used for the 6F pacing lead to prevent peri-lead bleeding from liver parenchyma. Post procedural ultrasonogram did not show any evidence of any local liver injury, hematoma or hepatic vein thrombosis.

### 2.2. Hybrid peratrial pacing

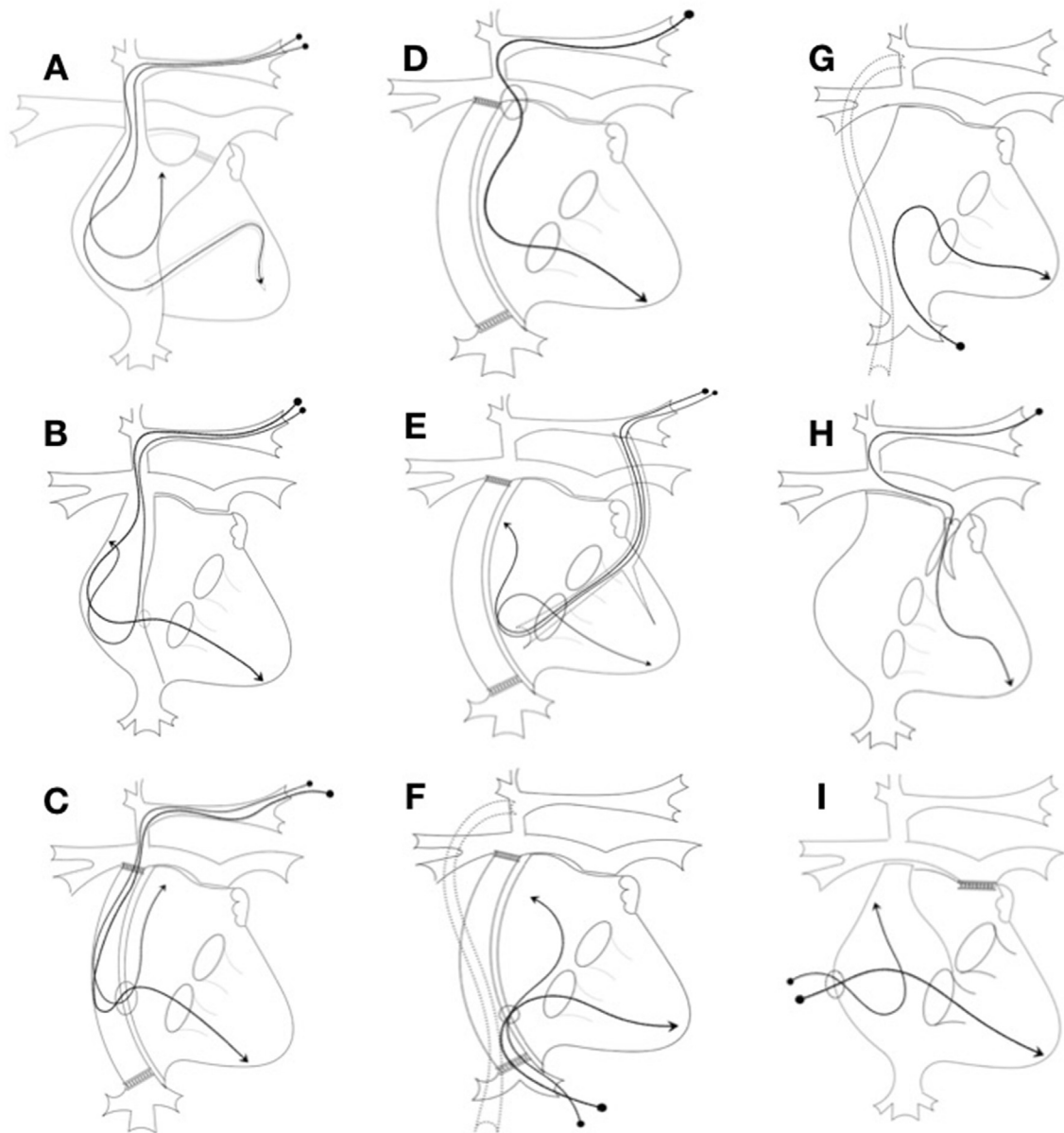
A 23-year-old female with Ebstein's anomaly of tricuspid valve underwent surgical tricuspid valve annuloplasty and Glenn anastomosis from right superior vena cava to right pulmonary artery. The atrial septal defect left open due to hypoplasia of the functional right ventricle showed right to left shunt, which increased the risk of systemic thromboembolism in the presence of an endocardial pacing lead. An epicardial dual chamber pacing was instituted for preexistent sinus nodal and atrioventricular nodal disease. In spite of normal pacing parameters during implantation, she developed symptomatic diaphragmatic pacing from both the leads after three months. Since extensive dissections were needed for lead revision through the previous surgical adhesions, peratrial endocardial pacing through a minimal access right anterolateral thoracotomy was performed after placing two purse string sutures on the right atrial free wall. After introducing two sheaths through the sutures, screw-in endocardial leads were secured in right atrial appendage and right ventricle (Fig. 3). On a follow up of eight years, pacing thresholds were stable and she had a recent pulse generator change through a small rectus sheath incision.

### 2.3. Endocardial leads through fenestration

Fenestrations are often surgically created during Fontan surgery as a pop off for the pulmonary circulation to improve the preload to the ventricles. A 14-year-old male who underwent fenestrated extracardiac conduit Fontan surgery for double outlet right ventricle, hypoplastic left ventricle, presented many years later with syncopal episodes secondary to progressive high-grade atrioventricular nodal block. A contrast angiogram in the conduit delineated the location of the fenestration (Fig. 4). As it was difficult to steer the pacing leads through the medially placed fenestration into the atrium and ventricle, a steerable telescopic method using a peel away coronary sinus catheter (Selectra BIO2-45, Biotronic, Berlin, Germany) and a steerable decapolar electrophysiological catheter (Inquiry, St Jude medical, Irvine, CA) facilitated the entry into the cardiac chambers through the fenestration. As the fenestration was not large enough to accommodate two coronary sinus sheaths, it was balloon dilated initially with an 8 mm Z-med (NuMED Inc, Hopkinton, NY) balloon. The steerable decapolar catheter also facilitated manipulation of the coronary sinus sheaths into the atrial appendage and both the leads were screwed into the ventricular apex and appendage respectively.

### 2.4. Endocardial leads through small venous collaterals

Following venous diversion of superior and inferior vena cava into a high-pressure pulmonary arterial tree, venous collaterals develop from them to decompress the caval veins to either the coronary sinus or the pulmonary veins. An eleven-year-old male child underwent extracardiac conduit Fontan for univentricular heart in 2008 followed by epicardial permanent pacemaker implantation in 2012 for heart block. The high pacing thresholds resulted in premature battery depletion within 6 years. A small persistent tortuous left superior vena cava draining into the coronary sinus was identified as a venous pop off from the innominate vein (Fig. 5). The entry into this tortuous venous channel by two leads was facilitated by a similar telescopic system described in 2.3. The same steerable system further facilitated the sheath to take a more than 360° turn from the coronary sinus to enter the atrioventricular valve and screw-in an endocardial lead in the ventricular apex.

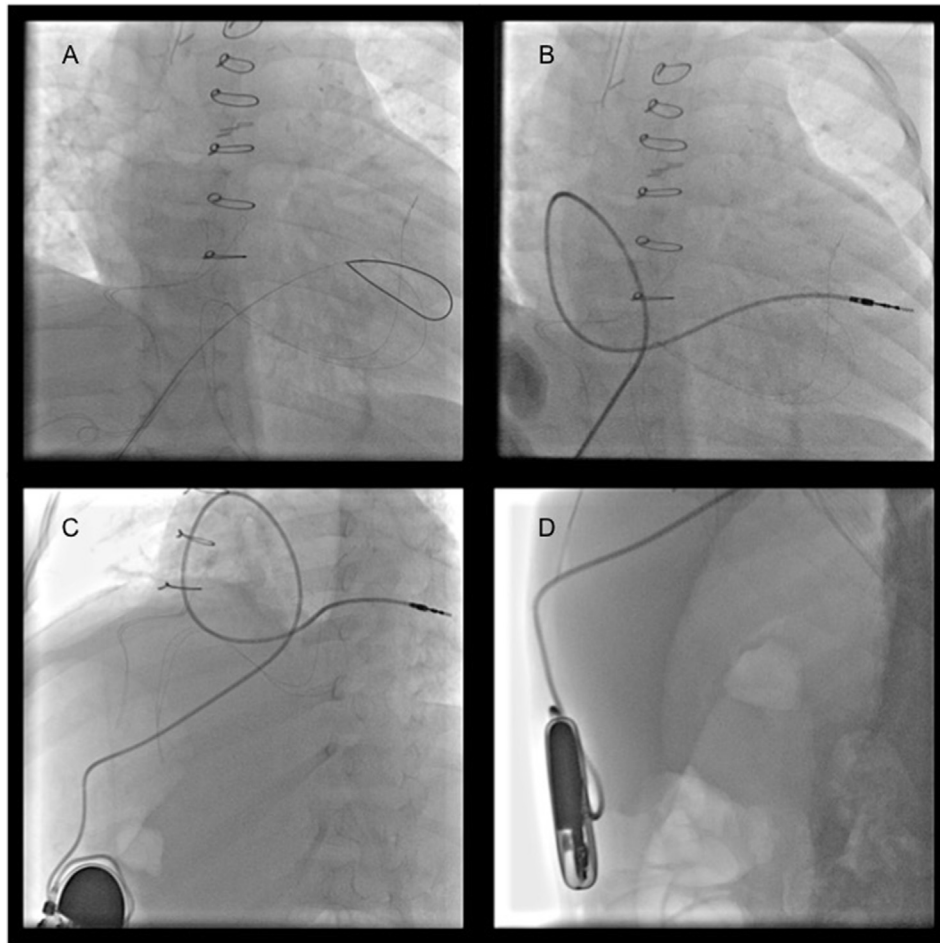


**Fig. 1.** Graphic illustration of different methods of endocardial pacing. A: Atrial lead in right atrial appendage and ventricular lead through the coronary sinus in atriopulmonary Fontan; B: Atrial lead in lateral atrial wall and ventricular lead through fenestration in lateral tunnel Fontan; C: Both atrial and ventricular lead through fenestration in extracardiac conduit Fontan; D: Ventricular lead through a fenestration created between floor of right pulmonary artery and atrium with a Brockenbrough needle in catheterization laboratory; E: Atrial and ventricular lead through venous collaterals after Fontan; F: Transhepatic leads through fenestration in extracardiac conduit Fontan following Kawashima shunt; G: Transhepatic lead through hepatic vein following total cavopulmonary Kawashima shunt; H: Ventricular lead through narrowed pulmonary valve in pulsatile Glenn shunt; I: Peratrial atrial and ventricular lead through a pursestring suture in right atrial wall.

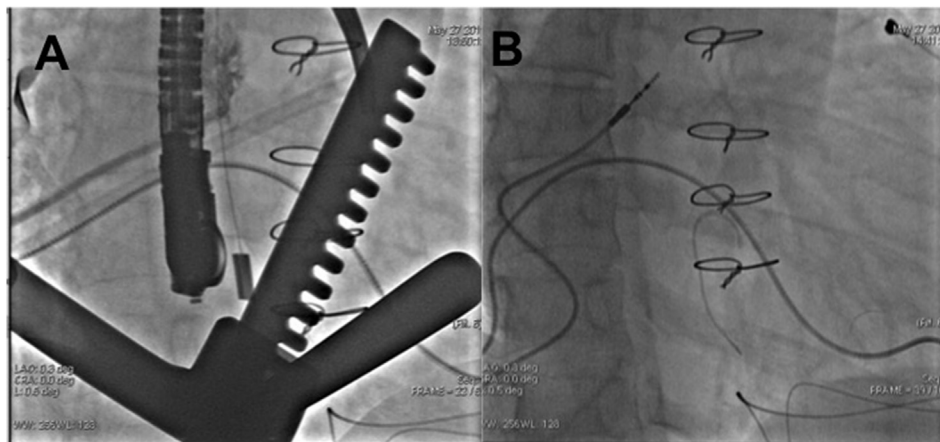
### 2.5. Combined epicardial and endocardial approach

The endocardial approach is often employed after a failed epicardial pacing, as the latter is always the initial preferred approach in young children. A six-month-old child diagnosed to have large inlet and additional multiple muscular ventricular septal defects and hypoplastic tricuspid valve was initially palliated with pulmonary artery banding followed by corrective surgery. An epicardial dual chamber pacemaker was implanted for a postoperative complete heart block. When the epicardial ventricular lead thresholds progressively increased to result in total ventricular pacing failure, it was

replaced by a ventricular endocardial lead from the left subclavian vein. As the previous atrial epicardial buttoned leads connected to a rectus sheath pulse generator still had a normal sensing and pacing function, the new implanted endocardial screw-in lead through the left subclavian vein was tunneled through the subcutaneous tissue in the chest wall to the same pulse generator in the epigastrium (Fig. 6). Even though this case scenario does not truly represent a post Glenn situation, it is added to highlight the space constraints in infants and very young children where lack of space for a pulse generator in the subclavian pocket will force the operator to choose a surgical epicardial option (see Fig. 7).



**Fig. 2.** A percutaneous venous access through the hepatic vein into the atria using a 0.025" guide wire(A) aided placement of a ventricular screw-in lead with a redundant loop in the atrium(B) and subsequently connected to a pulse generator(C) fixed in a subrectus pocket(D).



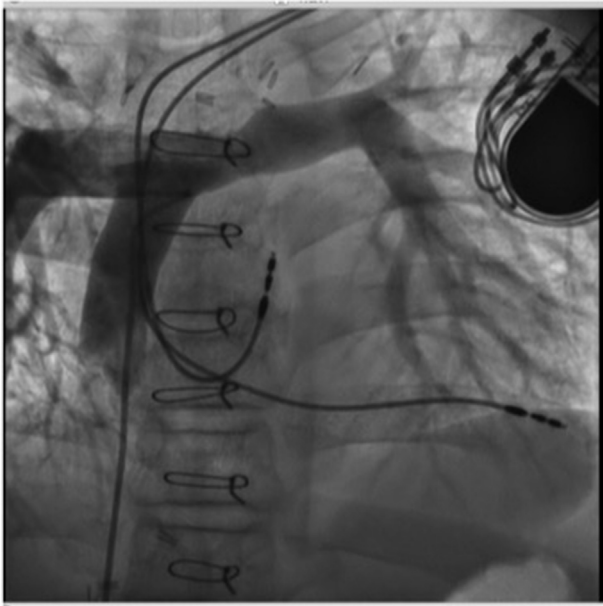
**Fig. 3.** A minimal access right anterolateral thoracotomy enabled placement of two peratrial sheaths(A) that enabled placement of atrial and ventricular screw-in leads. (B).

### 3. Discussion

#### 3.1. How common is the need for pacing after Fontan surgery?

Fontan surgery has substantially improved survival of patients with univentricular heart. 5–41% of survivors need permanent pacing at 15–20 years following surgery, making this the most

common intervention following Fontan surgery [14]. Atrial pacing for sinus node dysfunction has been reported in 9–23% following Fontan operation [1,2,15]. Even though atrioventricular nodal conduction abnormalities are less common than sinus nodal dysfunction, they follow antiarrhythmic use for the frequent atrial tachyarrhythmias and increase the need for ventricular pacing in many patients [1]. Certain morphologies of univentricular hearts



**Fig. 4.** The atrial and ventricular leads were advanced through a fenestrations using a steerable decapolar catheter telescoped in a coronary sinus peel-away sheath and finally screwed into the atrial appendage and ventricular apex respectively. (D).

associated with heterotaxy, common atrioventricular valves, atrioventricular discordance and some single ventricles are more prone for development of complete heart block.

### 3.2. Epicardial pacing as first choice

The venacaval diversion from the cardiac chambers following Fontan surgery makes epicardial pacing the preferred method. The extensive scarring following multiple thoracotomies associated with this surgery offers challenges to find healthy atrial and ventricular epicardium for lead placement [5]. The high epicardial pacing thresholds result in early battery depletion and generator replacement [6]. As lead failure occurs in 17–40% at 5–10 years following atrial epicardial leads, frequent lead and battery changes may be warranted in these patients resulting in high morbidity and costs [7]. The poor performance of epicardial leads justifies the extended indication of transvenous endocardial lead placement in univentricular hearts [16].

### 3.3. Why not endocardial pacing in Fontan patients?

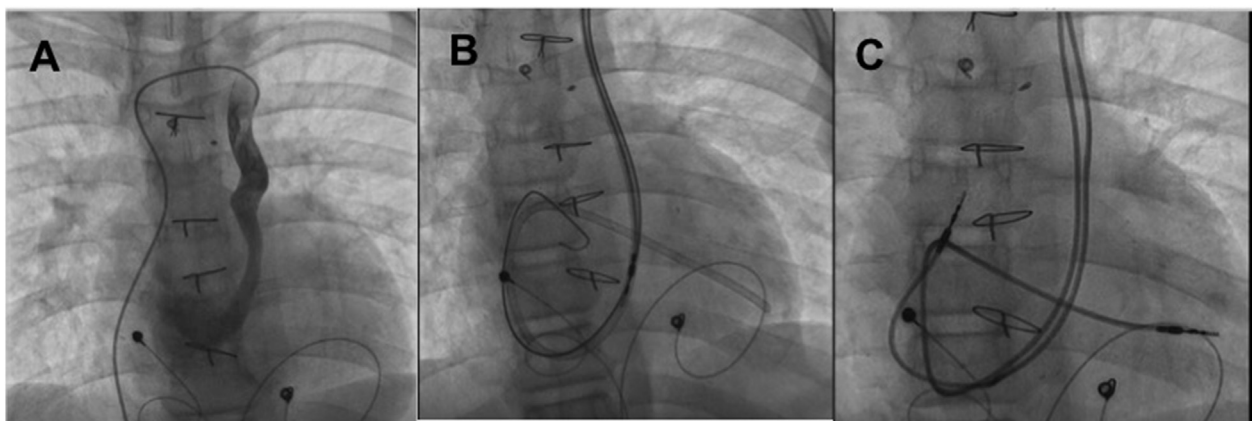
Endocardial pacing has excellent lead function and longevity owing to lower thresholds but has concerns of systemic thromboembolism, Fontan pathway obstruction by transvenous leads, risk of systemic atrioventricular valve regurgitation and infective endocarditis [10]. Moreover, venous diversion removes an easy access to the heart from the subclavian veins. The small size of the patients and attendant small sizes of the veins poses a risk of venous occlusions. There is also a higher risk of pulse generator extrusion from the pocket due to skin necrosis in the thin chest walled young children [11].

### 3.4. Systemic thromboembolism

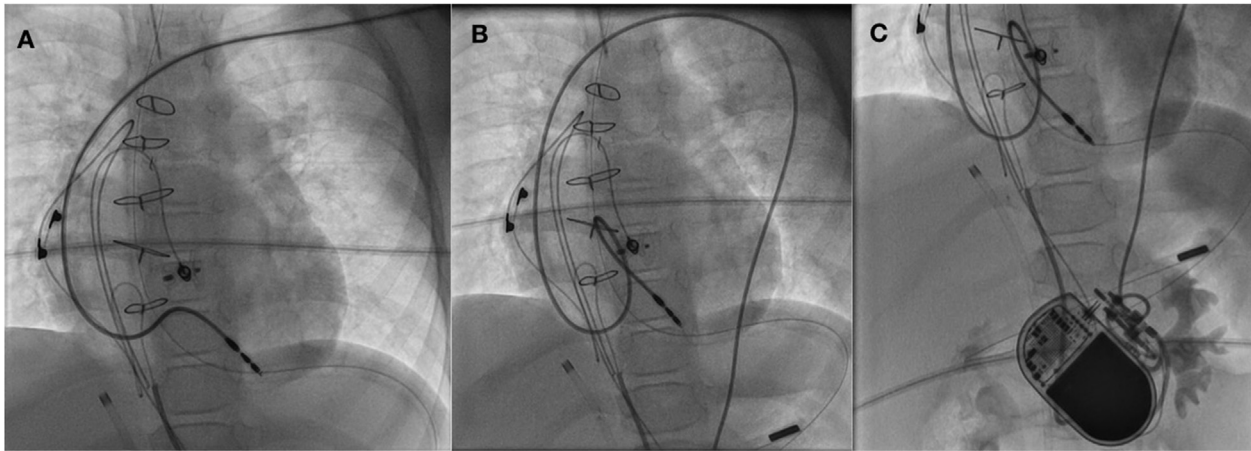
The risk of systemic thromboembolism from a right heart endocardial lead due to paradoxical right to left flows in patients with intracardiac shunts is 0.5–0.7 per patient-year. A pacing lead placed inadvertently in the left heart doubles this risk [12]. Fontan circulation carries the risk of increased thrombogenicity for reasons mentioned in section 1.3 [10,11]. Institutional preferences vary regarding anticoagulant management of Fontan circulation. Endocardial lead in Fontan circulation is likely to further increase the risks, though no trials have addressed this question. Systemic thromboembolic events in inadvertently placed left heart endocardial leads are effectively prevented with optimal anticoagulation [8,13]. Both epicardial and endocardial pacing in Fontan circulation are associated with thrombotic events indicating that the risk is related to the underlying non-pulsatile sluggish circulation rather than due to the pacing leads [17]. Many institutions prefer anticoagulation in every patient with Fontan circulation with a target international normalized ratio of 2–2.5. A multicenter ALSYNC registry currently assesses feasibility and benefit with elective left ventricular endocardial pacing along with adequate anticoagulation [18].

### 3.5. Risk of atrioventricular valve regurgitation

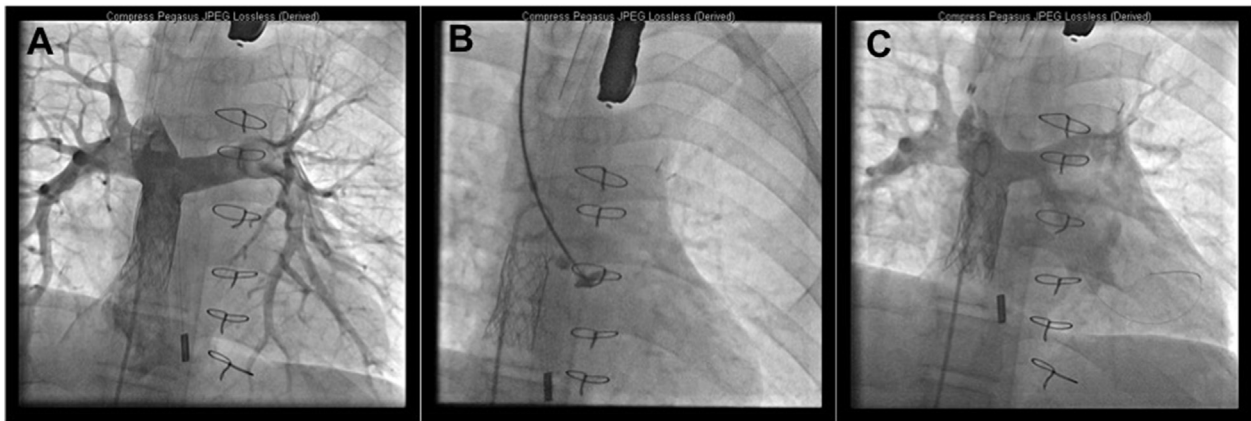
Endocardial transvenous pacing lead passing through tricuspid valve may lead to fibrosis or perforation of the leaflets, immobility due to leaflet impingement, trapping of the chordae and progressive regurgitation in few patients [19]. As pacing is associated with a 2.3 fold increased risk of development of significant tricuspid regurgitation and 1.6 fold increased risk of mortality due to the same, ongoing clinical trials aim to study merits of avoiding



**Fig. 5.** Following an angiogram to delineate the small tortuous venous collateral from left innominate vein to the coronary sinus(A), a steerable telescopic system guided placement of two endocardial leads(B) into the final position(C) to achieve dual chamber synchronous pacing.



**Fig. 6.** Combined epicardial and endocardial pacing. When epicardial ventricular lead failed due to lead fracture, it was replaced by an endocardial lead(A) placed inserted from left subclavian vein(B) and tunneled into the epigastric subrectus pocket (C)through the chest wall.



**Fig. 7.** When an extracardiac conduit Fontan had no fenestration(A), it was created with a transjugular Brockenbrough needle(B) and stented to create a fenestration from floor of right pulmonary artery to roof of the atrium(C).

transvenous leads in appropriate subjects [20,21]. In patients with Fontan circulation, the lead traverses through the systemic atrioventricular valve. Hence a significant regurgitation of this valve may impact the Fontan circulation significantly on long-term follow-up [22]. In patients with pulsatile Glenn shunt or persistent antegrade flows through the pulmonary outflow tract, the narrowed pulmonary valve can be crossed with a sheath from the subclavian vein for placing a ventricular pacing lead which avoids lead passage through the atrioventricular valves [23]. Atrioventricular lead is also avoided by a coronary sinus lead, when coronary sinus ostia is accessible from endocardial route as in older atriopulmonary Fontan surgeries [24].

### 3.6. Risk of endocarditis

Infections of pacing devices and leads has a reported incidence of 2.1–6.2% [25–27]. Complex larger devices like defibrillators and resynchronizing devices in children and young adults are more frequently associated with endocarditis [28]. Infections were the reason for 8.8% of all lead extractions [29]. There are no specific reports that suggest a higher incidence of pacing lead infections in patients with Fontan circulation. However, repeated device revisions and oral anticoagulation inherent in pacing following Fontan surgery is associated with higher incidence of endocarditis [30].

### 3.7. Patient size constraints

Dual chamber pacing provides atrioventricular synchrony and improves ventricular preload. The preload deprived Fontan circulation warrants dual pacing if there is bradyarrhythmia with atrioventricular dyssynchrony [2]. However a single chamber pacing is adopted in very small patients. In sick sinus syndrome following Fontan surgery, atrial pacing alone may be sufficient in small patients with normal atrioventricular nodal conduction [7]. A second additional lead and larger battery in a dual chamber pacing is associated with higher incidence of venous occlusions, pulse generator extrusions, skin necrosis and infections [31].

### 3.8. Pacing in the presence of venous occlusions and anomalies

Fontan circulation diverts venacava away from the cardiac chambers. When superior vena cava is occluded following Senning or Mustard surgery, stenting of the vein provides the access for an endocardial lead [32]. Hybrid peratrial approach described in section 2.2 has also been advocated in such patients [33,34]. Peratrial approach is safe, less morbid, offers low pacing threshold and can be applied widely with few contraindications. Peratrial lead placement has the additional advantage to allow somatic growth by creating a smaller redundant atrial lead loop [34]. Venous

collaterals representing remnants of persistent left superior vena cava may develop after Fontan surgery and allow access to the coronary sinus. Manipulating leads as described in section 2.4 may permit placement of atrial and ventricular leads in these patients, even though it may prove to be challenging [35]. Placement of pacing leads through the inferior vena cava in patients after Glenn shunt risks atrial lead dislodgement in 20% of patients and venacaval thrombosis/phlebitis [36,37].

### 3.9. Transhepatic lead placement

Patients with left isomerism, azygos continuation of interrupted inferior venacava and univentricular hearts are palliated with Kawashima shunt, where the superior venacava is anastomosed to the pulmonary artery. Following this surgery, the only direct access to the ventricles is through the hepatic veins. Section 2.1 and other reports describe the technique of transhepatic endocardial lead placement into the ventricles [38,39]. This transhepatic route is avoided in active liver disease, liver abscess or peritonitis.

### 3.10. Fenestrations in Fontan circuit to gain access into the heart

Fenestrations are surgically created during Fontan surgery to augment the preload to the ventricles and create a pop off for the pulmonary circulation [40]. The fenestrations in lateral tunnel Fontan is used with relative ease to gain access to the heart, but acute angulations inherent in fenestrations within an extracardiac polytetrafluoroethylene conduits challenge an entry by two pacing leads. Innovative steerable electrophysiology catheters and coronary sinus sheaths aid this maneuver as described in Section 2.3. In patients with no fenestration, the undersurface of the pulmonary artery can be punctured using Brockenbrough needle or radio-frequency to gain entry into the atrium (Fig. 6). These fenestrations can then be used to place endocardial leads [41–43].

### 3.11. Older modifications of Fontan surgery

Extracardiac conduit Fontan surgery is preferred recently as the method of choice due to reduced thrombogenicity and least arrhythmogenicity. Among the previous modifications as in Hemifontan procedure, the superior vena cava-right atrial junction sleeve which is remaining in direct continuity with the venous system can be paced [44]. In the atriopulmonary Fontan surgery which is almost abandoned today, both the entire right atrial appendage and coronary sinus ostium are available for placing atrial and left ventricular leads [45–48]. In the intracardiac lateral tunnel Fontan surgery with creation of an intra-atrial baffle through a part of the right atrial wall to direct the inferior vena caval blood to the pulmonary circulation, the atrial wall can be used for atrial pacing and the baffle fenestrated for gaining a ventricular lead [17,41]. Another modification is extracardiac lateral tunnel surgery where a portion of the epicardium of the right atrium that forms a part of the wall of the baffle leading up to the pulmonary artery anastomosis is used for pacing [49,50]. However mapping the atrial tissues which could be paced effectively and avoiding fibrotic areas, avoiding phrenic nerve stimulation, securing leads in the most appropriate locations can be challenging.

## 4. Conclusions

The prolonged survival of patients with univentricular hearts after Fontan surgery and its recent modifications has led to a significant proportion of these survivors needing pacemaker implantation for various forms of bradyarrhythmias. Even though the method of choice, epicardial pacing fails in many due to high

thresholds, lead fractures and difficulties in repeated surgical revision of leads due to adhesions. Endocardial lead placement needs special techniques and unusual access due to the venacaval diversion away from the heart during this surgery. Compared to the previous modifications of Fontan surgeries that offered an easier site to pace the heart within the right atrium, the present modification of using an extracardiac synthetic conduit for diversion of inferior venacava does not allow any native atrial tissue around the conduit. Endocardial leads have to be inserted through venous collaterals, hepatic veins, pulmonary valve, fenestrations created in surgery or catheterization laboratory or through the right atrial wall. The different access and varied techniques have created new options for pacing the univentricular heart for the bradyarrhythmias. The higher thrombogenicity of Fontan circulation may be compounded by endocardial transvenous leads and warrants adequate systemic anticoagulation, which prevents the risks of systemic thromboembolism. Other problems associated with endocardial pacing namely atrioventricular valve regurgitations, endocarditis are prevented by meticulous safe techniques.

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## Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ipej.2018.11.013>.

## Declaration of interest

None.

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None.

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