*Cardiology in the Young*

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# Persistent left superior vena cava accompanying repaired tetralogy of Fallot: Does it pose a challenge for device implantation?

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**Brief Report**

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**Abstract**

Persistent left superior vena cava is a thoracic venous return anomaly. Tetralogy of Fallot is one of the most common congenital anomalies seen with persistent left superior vena cava. We are presenting a successful cardiac resynchronisation therapy device implantation in a patient with repaired tetralogy of Fallot and persistent left superior vena cava combination which has not been previously reported in the literature.

## Case

A 62-year-old male patient with repaired tetralogy of Fallot and persistent left superior vena cava presented with ongoing dyspnea despite optimal medical treatment (NYHA class III). At the time of operation of tetralogy of Fallot, the patient was 17 years old. In the patient's history, there was no need for pulmonary valve replacement.

Electrocardiogram showed atrial fibrillation, right bundle branch block, and left anterior hemiblock. The QRS duration was 186 ms (Fig 1). Transthoracic echocardiography showed biventricular systolic dysfunction (left ventricular ejection fraction: 32%, tricuspid annular plane systolic excursion: 10.2 mm). He had a dilated coronary sinus (31.6 mm). In transthoracic echocardiography, there was no right ventricular outflow tract obstruction (valvular or subvalvular) but only mild pulmonary regurgitation was present. The residual ventricular septal defect was not observed. The persistent left superior vena cava was detected by contrast echocardiography. A cardiac resynchronisation therapy device with defibrillator implantation was decided. Venography was performed through left subclavian vein access. The existence of persistent left superior vena cava was verified. An active fixation lead was preferred as the defibrillation electrode. This lead was advanced through the dilated coronary sinus to the right atrium.

A stylet with around angle of 110 degrees was folded in the right atrium and by advancing from here lead was finally fixed into the right ventricle. The coronary sinus branch was identified by light opaque injections from an angled lateral branch catheter in the access catheter. The support of the access catheter was increased by leaning its folding point on the opposite wall of the dilated coronary sinus. The lead of the quadripolar coronary sinus was successfully advanced through extra support wire. The atrial lead was not implanted because of permanent atrial fibrillation. After a cardiac resynchronisation therapy device with defibrillator implantation, the QRS width was 166 ms (Fig 2). The patient's symptoms improved after the implantation (NYHA class II). Also, after 6 months of follow-up, the functional status remained the same.

## Discussion

The long-term survival rate of repaired tetralogy of Fallot patients is well. The number of repaired tetralogy of Fallot patients is increasing in the adult population.[1] Beginning from the early post-operative period, patients present many conduction abnormalities (often right bundle branch block)[2] and these disorders tend to progress over time. Right ventricular[3] and left ventricular[4] systolic dysfunction can also be presented in repaired tetralogy of Fallot patients. In these situations, device implantation may be required. It has been reported that in short to medium term, significant clinical improvement has been observed following cardiac resynchronisation therapy in patients with operative tetralogy of Fallot and moderate or severely decreased left ventricular systolic functions.[5] A cardiac resynchronisation therapy device implantation in a patient with repaired tetralogy of Fallot and persistent left superior vena cava combination has not been previously reported in the literature. Persistent left superior vena cava was reported in 5.9% of tetralogy of Fallot patients.[6] In patients with persistent left superior vena cava, cardiac resynchronisation therapy device implantation can be performed successfully through the persistent left superior vena cava despite several anatomical difficulties. We also performed successful implantation with the methods mentioned above.



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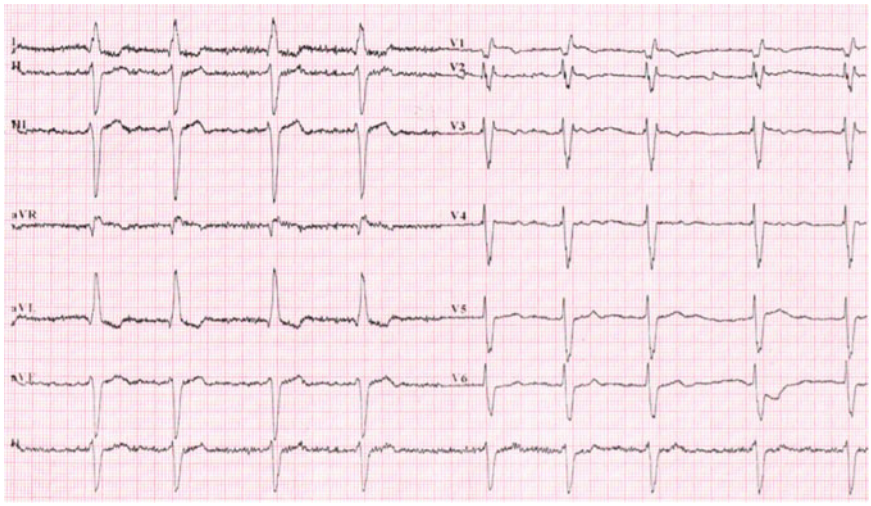

**Figure 1.** Electrocardiogram with standard lead positioning before implantation.

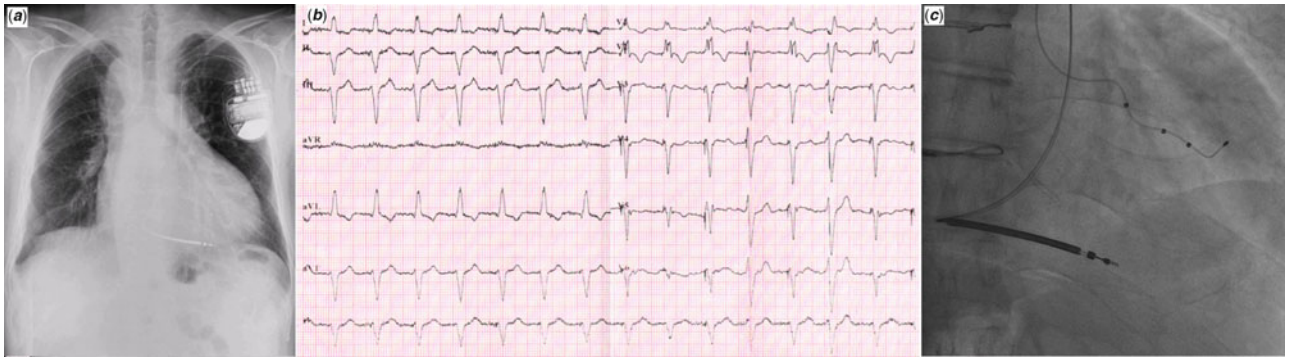

**Figure 2.** Biventricular lead position after implantation (Panel a and c). Electrocardiogram after implantation (Panel b).

Surgical techniques for the correction of tetralogy of Fallot have improved dramatically since the first operation was performed in 1955 and patients are living well into adulthood. With increased survival, increased morbidity secondary to long-term complications of tetralogy of Fallot is seen. So, it is important for the management of long-term outcomes of repaired tetralogy of Fallot patients.

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**Conflicts of Interest.** None.

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