Pacemaker Implantation with Absent Right Superior Vena Cava

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Abstract
Persistent left superior vena cava with absent right superior vena cava is a rare venous congenital malformation (ARSVC). This is the first report of images of two pacemakers implantation into the patient with ARSVC.

Introduction
A persistent left superior vena cava (PLSVC) with an absent right superior vena cava (ARSVC) is a very rare anomaly. A previous report demonstrated that rhythm disturbances such as sinus node dysfunction and atrioventricular block have been seen in the patient with this anomaly. The anomaly must present difficulties in right ventricular pacemaker lead insertion. This case report demonstrates that two pacemakers implantation into the patient with ARSVC and combination of an atrial single-chamber (AAI) pacemaker and a leadless pacemaker can work as dual chamber (DDD) pacing system in our patient.

Case presentation
A 75-year-old man underwent pacemaker implantation because of symptomatic sick sinus syndrome (SSS). Preoperative venography confirmed an ARSVC and PLSVC. Right ventricular pacing was considered difficult, and he was implanted with an AAI pacemaker lead through PLSVC for SSS. Two years after operation, he was admitted to our hospital for symptomatic complete atioventricular block. Three-dimensional CT scan also revealed an ARSVC and a PLSVC (Figure 1A), and we implanted a leadless pacemaker (LP) in right ventricle (RV) via the right femoral vein (Figure 1B, arrow). We checked LP could follow not only the autologous atrial wave, but also the wave triggered by atrial pacing, indicating combination of AAI pacemaker and LP can work as dual chamber pacing system in our patient (Figure 1C). He has been free of symptoms since the LP implantation.

Discussion
An ARSVC with a PLSVC without any other cardiac anomalies is an extremely rare anomaly. Because the lack of symptoms, this malformation is often detected fortuitously when patients undergo pacemaker implantation. The condition should present difficulties during RV pacemaker lead insertion. In addition to the previous AAI pacemaker, we implanted a LP in RV and the combination of these pacemakers has worked as a dual chamber pacemaker in our patient. We should keep this rare anomaly in mind, especially when we insert RV pacemaker lead.
Novel Teaching Points

1. An absent right superior vena cava (ARSVC) with a persistent left superior vena cava without any other cardiac anomalies is an extremely rare anomaly.

2. We implanted a leadless pacemaker in right ventricle (RV) and the combination of these pacemakers has worked as a dual chamber pacemaker in our patient.

3. We should keep this rare anomaly, ARSVC, in mind, especially when we insert RV pacemaker lead.

References


**Figure 1:** Three-dimensional CT scan revealed an absent right superior vena cava with a persistent left superior vena cava (A), and we implanted a leadless pacemaker (LP) in right ventricle via the right femoral vein (B, arrow). LP could follow not only the autologous atrial wave, but also the wave triggered by atrial pacing, indicating combination of AAI pacemaker and LP can work as dual chamber pacing system in our patient (C).

**Declarations**

**Conflict of interest:** The authors have no conflicts of interest to declare.

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