CASE REPORT

Cardiac resynchronization therapy in a case with single ventricle and concommitant noncompaction cardiomyopathy

Kardiyak resenkronizasyon tedavisi ile başarılı tedavi edilen nonkompaksiyon kardiyomiyopatili tek ventrikül

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Summary– A 32-year-old male patient was admitted to the hospital with syncope. An electrocardiogram revealed complete atrioventricular (AV) block and a right bundle branch block with a QRS duration of 218 milliseconds. The heart rate was 40 beats/minute. Echocardiography revealed that both AV valves opened to a single ventricle as well as noncompaction of the myocardium. Due to New York Heart Association class III heart failure, cardiac resynchronization therapy with a defibrillator device (CRT-D) was performed. After the implantation, the electrocardiography QRS duration was reduced to 183 ms. To our knowledge, this was the first reported case of CRT-D implantation in a patient with a noncompacted single ventricle and complete AV block.

Özet– Otuz altı yaşında erkek hasta senkop ile hastaneye yatırıldı. Elektrokardiyografide QRS süresi 218 msn olan atriyoventriküler (AV) blok ve sağ dal bloğu tespit edildi. Kalp hızı 40 atım/dk olarak görüldü. Ekokardiyografide nonkompaksiyon tek ventrikül olduğu görüldü. Hastanın kalp yetersizliği New York Kalp Cemiyeti sınıf III olması nedeniyle, defibrilatör cihazı ile kardiyak resenkronizasyon tedavisi (CRT-D) planlandı. İmplantasyon sonrası kalp yetersizliği geriledi ve QRS süresi 183 ms olarak tespit edildi. Bu olguyu litaratürde, nonkompaksiyon tek ventrikül ve AV tam bloğu olan ilk CRT-D implantasyon olgusu olması sebebi ile sunuyoruz.

Single ventricle defects are rare; they comprise 1.5% of all congenital heart diseases (CHD). The primary symptoms are dyspnea and cyanosis. Frequently, a subvalvular or valvular outflow tract obstruction to either great artery is present.[1] The related causes of death can include heart failure, dysrhythmia, pulmonary embolism, thrombosis, and abscesses.[1] It is not uncommon for these patients to have a 17–30% atrioventricular (AV) block as a result of morphologically different AV node and His bundle placement.[2] This case report describes the successful treatment of complete AV block and heart failure in a patient with a single ventricle defect using biventricular pacing.

Abbreviations:

AV	*Atrioventricular*
CHD	*Congential heart disease*
CRT	*Cardiac resynchronization therapy*
CRT-D	*CRT with defibrillator implantation*
CS	*Coronary sinus*
CT	*Computed tomography*
ECG	*Electrocardiogram*
LV	*Left ventricle*
ms	*Milliseconds*

CASE REPORT

A 32-year-old male was diagnosed with a single ventricle at the age of 17. No further medical follow-up occurred. The patient was admitted to the emergency department with syncope. An electrocardiogram revealed a complete AV block with a ventricular escape rhythm at a rate of 40 beats/minute, right bundle branch block morphology, and a QRS duration of 218 milliseconds (ms) (Fig. 1a). The patient reported shortness of breath upon even minimal effort. His functional capacity was assessed as New York Heart Association class III. Clubbing of the fingers was observed. Pulse oximeter measurement of oxygen satu-

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ration was 90%. The hematological and biochemical parameters were normal. Echocardiographic images revealed that both AV valves opened to a single ventricle and noncompaction of the myocardium (Fig. 1c). The ejection fraction of the common ventricle was measured at 30% using Simpson's method. Computed tomography (CT) angiography revealed that the aorta was anterior and to the left of the main pulmonary artery (Fig. 1d). L-loop transposition of the great arteries was diagnosed. Cardiac magnetic resonance imagery demonstrated noncompaction of the ventricular structure (Fig. 2a-c). Coronary angiography results were normal; the pulmonary artery systolic pressure and the aortic systolic pressure were nearly the same during catheterization. Angiotensin-converting enzyme inhibitor and aldosterone antagonist treatment was administered. Cardiac resynchronization therapy with a defibrillator (CRT-D) was performed. Pacing device implantation may be performed via epicardial or transvenous access in patients with complex CHD. There are no studies comparing these 2 methods in unoperated, single-ventricle patients. The transvenous route was preferred in this case due to the patient's reluctance to undergo surgery and to avoid possible complications of surgery. Standard practice at our institution is to implant the left ventricle (LV) pacing lead using left subclavian access via a transvenous approach, with the coronary sinus (CS) engaged using a combination of a no deflectable guiding catheter. CS angiography was performed after entering the CS with a left anterior oblique 30° view using the CS catheter. After selecting the appropriate branch, it was passed with a soft wire and the LV lead was positioned (Fig. 3a, b). A persistent left superior vena cava was not detected on angiography or CT. After a quadripolar LV lead (Quartet; St. Jude Medical, Inc., St. Paul,

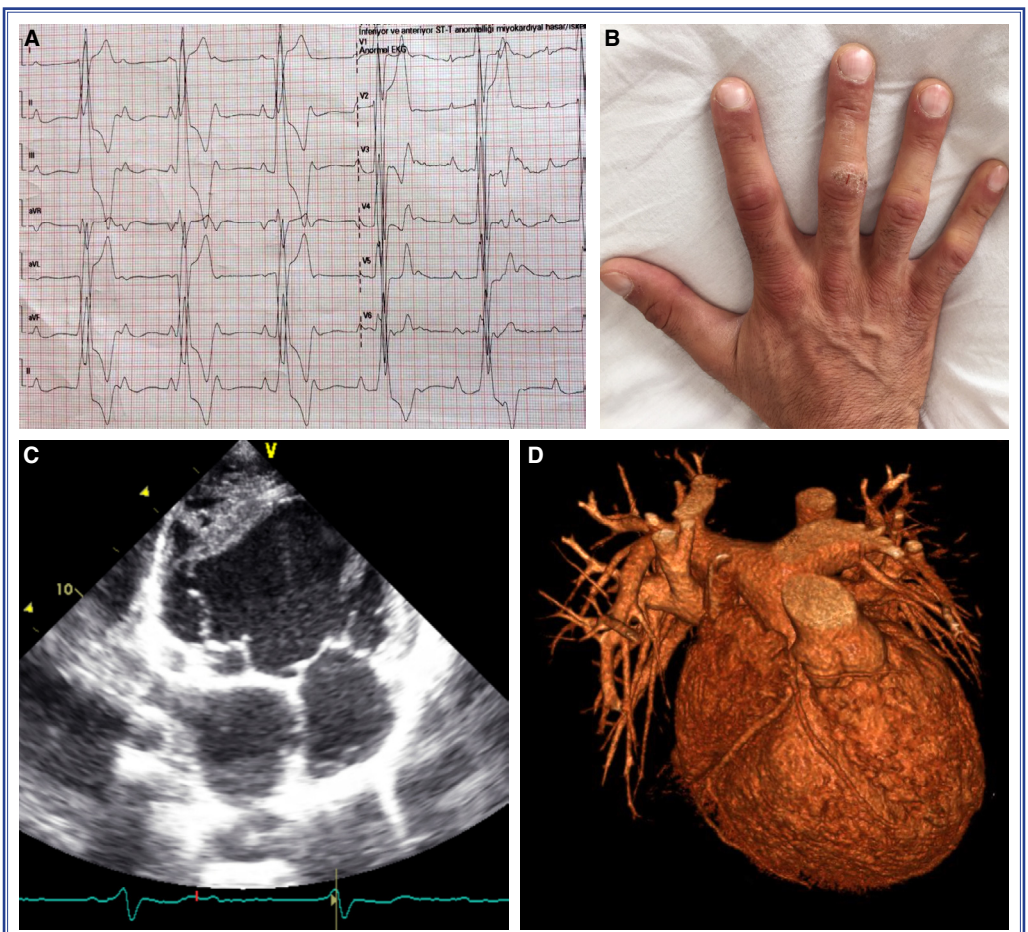

Figure 1. **(A)** Admission electrocardiogram showing complete atrioventricular block; **(B)** Clubbing can be seen on the patient's right hand; **(C)** Transthoracic echocardiography apical 4-chamber view showing that both atrioventricular valves opened to a single ventricle with noncompaction; **(D)** Computed tomography angiogram image.

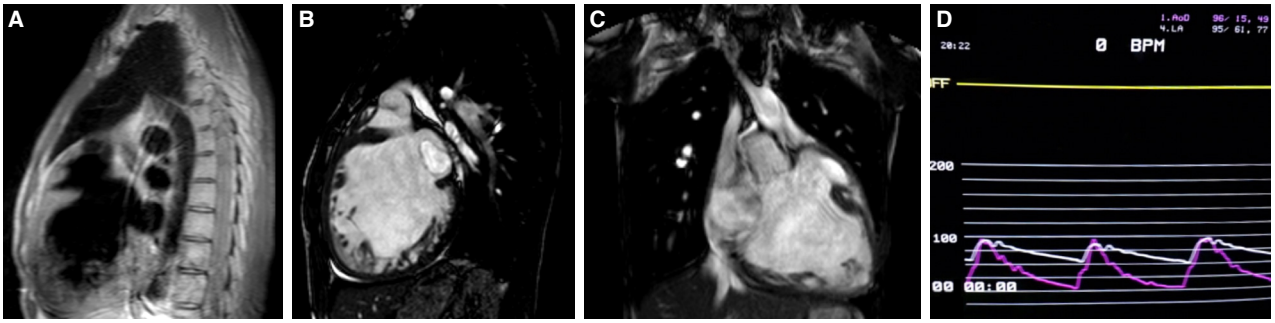

Figure 2. (A, B) Cardiac magnetic resonance (MR) images reveal ventricular noncompaction, frontal plane; **(C)** MR image of the single ventricular structure, sagittal plane; **(D)** Image of pulmonary artery pressure measurement with catheter.

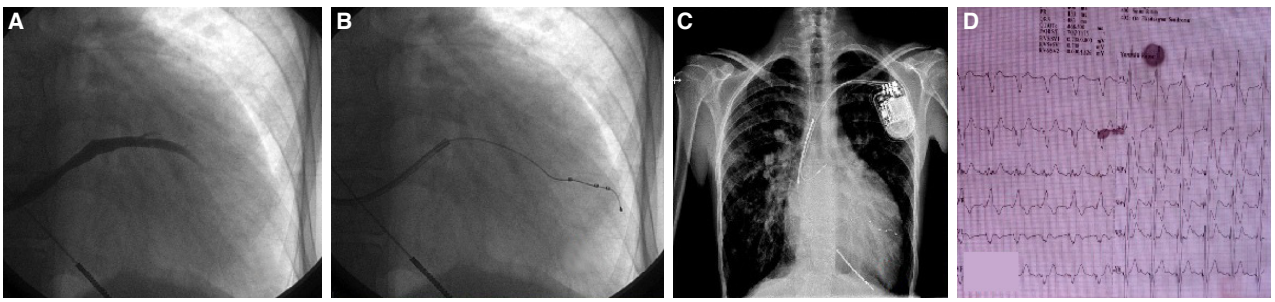

Figure 3. (A, B) The wire and left ventricular leads as seen in angiogram images; **(C)** Atrial, ventricular, and coronary sinus leads seen on a chest radiogram; **(D)** Electrocardiogram results after cardiac resynchronization therapy with defibrillator implantation showing that the QRS duration had decreased compared with the results seen upon admission.

MN, USA) was placed in the posterolateral branch of the CS, a CRT-D device (Quadra Assura MP; St. Jude Medical Inc., St. Paul, MN, USA) was implanted (Fig. 3c). The R wave was 10.5 mV and the excitation threshold was 1.7 mV in the LV lead measurements. The lead measurement was concluded with no phrenic nerve stimulation. Right atrial and ventricular leads were placed through the subclavian vein to the right atrium and ventricle.

Warfarin was added upon discharge from the hospital treatment for embolism. One month later, his functional capacity improved to New York Heart Association lass I. The QRS duration on an ECG was reduced to 183 ms (Fig. 3d). Echocardiography indicated that the ejection fraction of the common ventricle had improved to 40%. No cardiovascular mortality, hospitalization, increased symptoms of heart failure, arrhythmia, or cerebrovascular events were observed in the patient's 1-year follow-up. To our knowledge, this is the first reported CRT-D implantation case in an unoperated patient with a noncompacted single ventricle and complete AV block.

DISCUSSION

A single ventricle is a rare congenital heart malformation. If feasible, the goal is separation of systemic and pulmonary circulation by means of a total cavopulmonary connection during childhood. However, long-term results are still associated with significant mortality and morbidity caused by arrhythmia, venous congestion, thromboembolism, and ventricular failure.[3] In our case, the patient had complete AV block and class III heart failure.

Progressive heart failure is a major cause of death during late follow-up of patients with complex CHD. Within this population, there are several subgroups of patients according to the cardiac anatomy, including patients with a systemic LV, patients with a systemic right ventricle, and patients with a single ventricle. These different groups may have different responses to CRT.[4] CRT therapy has been shown to be successful in a heterogeneous group.[5] CRT is a useful treatment for adult patients with heart failure and selected forms of electrical synchronization with reduced ejec-

tion fraction, but patient selection and technical problems create significant difficulties in the adjustment for CHD.[6] There are often significant technical challenges in achieving CRT in cases of complex CHD, particularly via the endocardial route. Even if the CS is accessible, pacing of the systemic ventricle may not be achievable via this route. Outcome data for CRT in CHD are generally limited to small studies in the acute postoperative setting and longer-term follow-up studies with a variety of surrogate endpoints (e.g., 6-minute walk test, hospitalizations, quality of life, ventricular stroke volume/ejection fraction). In general, the systemic LV is more likely to respond to CRT than the systemic RV, and even in patients awaiting transplantation, the results are more promising in systemic LV cases.[7,8] Further CRT optimization, including refinement of AV and VV (interventricular) delay, may also be considered, but there is an absence of evidence and guidance regarding an optimal modality and settings in the CHD population.

The clinical effects of CRT on adult heart failure patients and the beneficial effects on LV functions have encouraged the use of this treatment in other populations, such as those with CHD and pediatric patients. In our case, CRT was applied as a result of adult CHD and advanced heart failure, and the symptoms of heart failure regressed and syncope completely resolved.

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Keywords: Cardiac resynchronization therapy with defibrillator implantation; noncompaction; single ventricle.

Anahtar sözcükler: Defibrilatör cihazı ile kardiyak resenkronizasyon tedavisi; non-compaction; tek ventrikül.