

CASE REPORT

# Cardiac Resynchronization in the Acute Phase of Decompensated Heart Failure in a Young Patient with Systemic Sclerosis

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## ABSTRACT

Cardiac resynchronization therapy is an essential tool for treating patients with heart failure in different types of dilatative cardiomyopathy with bundle branch block. This technique is widely used with significant benefits in terms of quality of life and effort tolerability along with optimized medical therapy. In spite of its benefits, several factors may influence its efficacy such as etiology, lead position, or device settings. In some cases, the anatomical variance of the coronary sinus could create technical difficulties for advancing the left ventricular lead. This case report describes a female patient presenting with decompensated heart failure, known with complicated sclerosis multiplex and a fibrous tissue in the coronary sinus, which created a critical obstruction leading to impossibility to advance the left ventricular lead. This case underlines the importance of appropriate imaging investigation for optimal interventional approach in these difficult cases.

**Keywords:** cardiac resynchronization, heart failure, systemic sclerosis, coronary sinus venoplasty, dilatative cardiomyopathy

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## INTRODUCTION

Heart failure (HF) is one of the most common causes of cardiovascular death and is associated with an increased need for repeated hospitalization.<sup>1,2</sup>

Dilated cardiomyopathy (DCM) represents one of the challenging causes of HF, with the highest prevalence in Central Europe and the highest mortality rate in Central and Eastern Europe. Cardiac resynchronization therapy (CRT) or multi-site ventricular pacing along with opti-

mized medical therapy can significantly improve the quality of life and may reduce short- and long-term mortality among patients with DCM and electrical dyssynchrony. Current guidelines establish a clear indication for CRT in DCM with a left ventricular ejection fraction (LVEF)  $\leq 35\%$  and a QRS larger than 150 ms for left bundle branch block (LBBB) morphology, in symptomatic patients despite optimized medical therapy. In case of QRS duration between 130 and 149 ms or larger but no LBBB morphology, CRT should be considered to improve the HF symptoms and

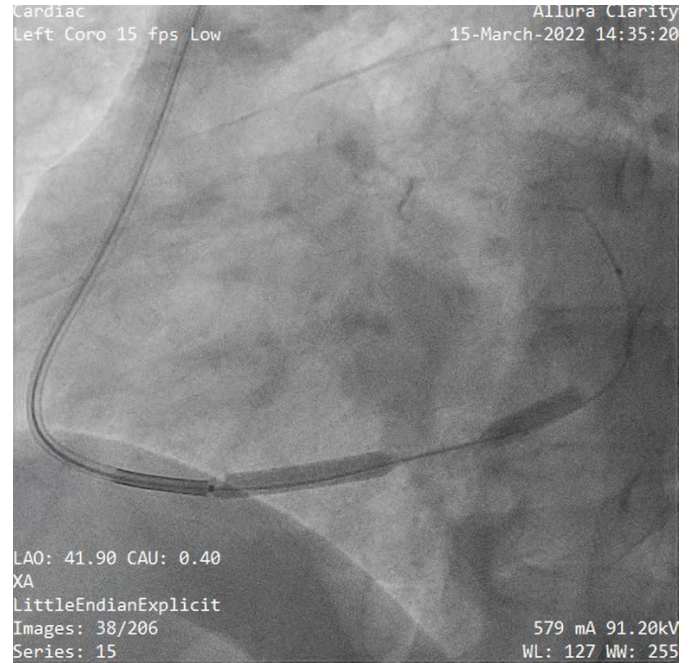
reduce mortality.<sup>3</sup> However, in some rare cases, anatomical variation of the coronary sinus (CS) could make the implantation procedure difficult or even impossible.

Several systemic inflammatory disorders have been incriminated in the etiology of HF, including systemic sclerosis (SSc), lupus etc. SSc is a rare, systemic heterogeneous connective tissue disease, often associated with micro- and macrovascular damage, as well as generalized fibrosis in organs due to excessive extracellular matrix deposition.<sup>4</sup> Cardiac involvement in SSc includes fibrosis, myositis, conduction abnormalities, coronary artery disease, and HF, with a clinical manifestation in up to 45% of cases.<sup>5</sup> Diffuse vascular fibrosis with contraction band and concentric intimal hypertrophy contribute to reducing the vasodilator reserve. This can manifest in reversible myocardial ischemia and myocardial abnormalities, which accelerate subendocardial myocardial fibrosis with HF, and systolic and diastolic dysfunction.

Here we present the case of a patient presenting with decompensated HF, in whom CRT was initiated, and coronary sinus venoplasty was required during the intervention because of occlusion of the proximal segment of the CS.

## CASE PRESENTATION

We present the case of a 60-year-old female patient, previously diagnosed with SSc associated with multiorgan manifestation – Sjögren's syndrome and Raynaud's syndrome. The patient presented moderate pericardial

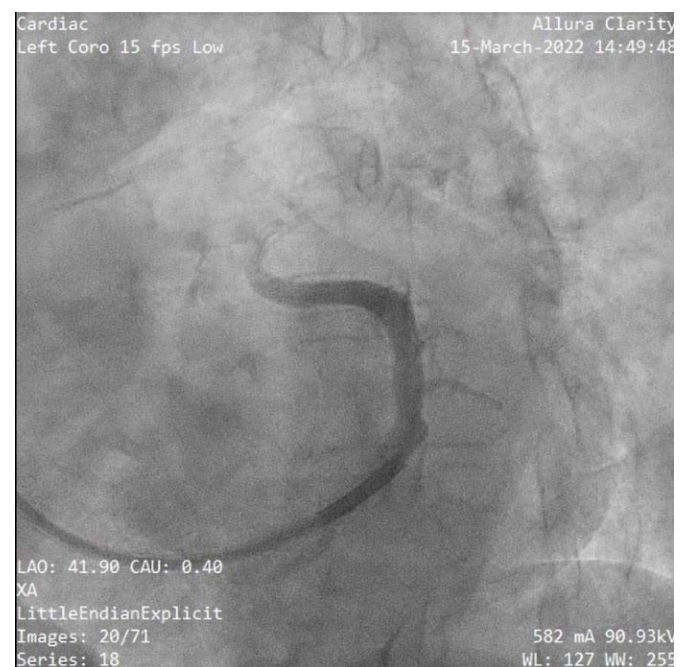


**FIGURE 1.** First venography with the proximal occlusion of the coronary sinus

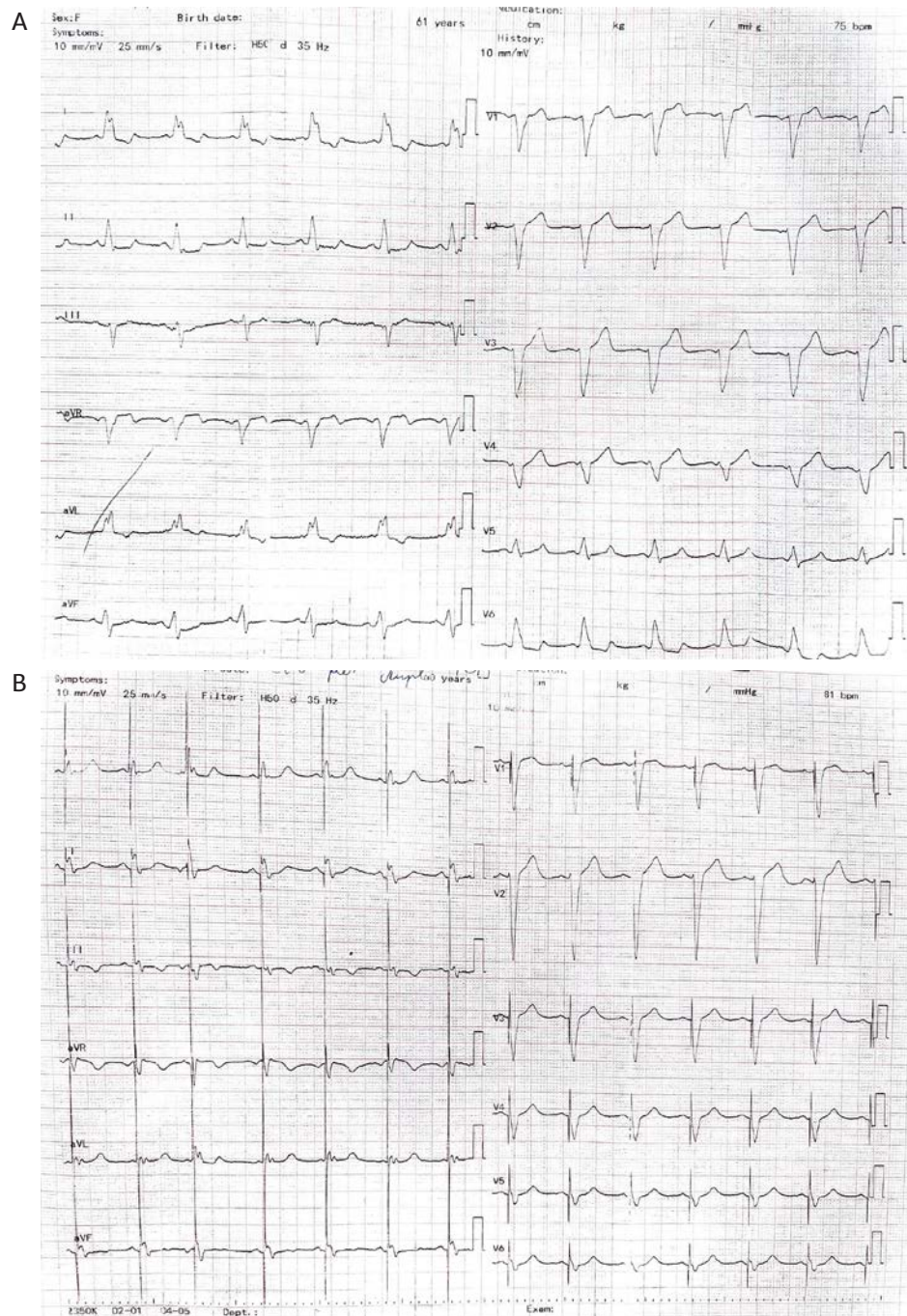
effusion, mitral and tricuspid valve insufficiency, carotid artery atheromatosis, and post-viral DCM with severe systolic dysfunction and LBBB, arterial hypertension, and HF. Coronary computed tomography angiography (CCTA) was performed to exclude an ischemic origin of DCM. At presentation, the patient had decompensated HF, fatigue, shortness of breath, diaphoresis, and hypertensive



**FIGURE 2.** Coronary sinus venoplasty



**FIGURE 3.** Venography after venoplasty



**FIGURE 4.** ECG before (A) and after (B) the CRT implantation

crisis. The symptoms occurred in spite of optimized oral medical therapy with beta-blockers, angiotensin receptor/neprilysin inhibitor (ARNI), and diuretics. Laboratory investigations revealed an elevated level of NT-proBNP (3,600 pg/ml) and mild microcytic hypochromic anemia. After admission, intravenous loop diuretics and iron sucrose perfusion were initiated, with slowly favorable evolution and decrease of NT-proBNP to 1,268 pg/ml after 48 h and 575 pg/ml at discharge.

Transthoracic echocardiography revealed a dilated left ventricle (LV) – 63/40 mm, severe systolic dysfunction, global hypokinesia of the LV, and an ejection fraction of 30%. ECG revealed sinus rhythm with LBBB and QRS duration of 162 ms.

The initial approach to CRT implantation consisted in the conventional method for preparation. First, a special sheet was inserted from the subclavian vein to facilitate the cannulation of the CS. After failure to insert the LV lead,



an Amplatz left guiding catheter was positioned in the CS to advance the catheter for a better venography and to visualize potential anatomical difficulties such as stenoses or valve persistence. Venography revealed an occlusion at 1 cm after the origin of the CS (Figure 1).

After venography, we decided to perform percutaneous coronary venoplasty using a 0.0014-inch hydrophilic guidewire that was advanced through the occlusion, deep in a collateral vein to achieve more stability. This was followed by balloon angioplasty in the proximal segment of the CS vein, using a peripheric semi-compliant 6 × 120 mm balloon (Abbott Armada Dilatation Catheter). Under fluoroscopic control, the balloon pressure was gradually increased until the stenosis has been dilated. After dilatation, control venography was performed with a permeabilized CS vein revealing an optimal side branch for the LV lead position (Figure 2, 3).

The venoplasty was followed by a control venography, and the LV lead was advanced in the anterolateral coronary vein to the anterolateral surface of the ventricle, with an optimal pacing position. The implantation was finalized with the implantation of the atrial and right ventricular leads.

The ECG showed a significantly shortened QRS, from 162 ms to 122 ms (Figure 4). The control echocardiography at discharge revealed an improved LV function (LVEF increased from 30% to 40%).

The one-month follow-up revealed a slightly ameliorated LV function, with a negative remodeling with respect to the LV diameter (from 63 mm to 58 mm), a slight improvement in mitral regurgitation, and an important improvement in symptoms.

The patient provided written informed consent allowing the publication of her data, and the institution where the patient had been admitted approved the publication of the case.

## DISCUSSION

CRT can play an important role in the treatment of HF with large QRS. There are four main mechanisms through which CRT can have an impact on the myocardium.<sup>6</sup> The first one is the correction of LV dyssynchrony, by incurring a delay in the contraction of the opposite wall using an extra stimulus of the LV lead implanted on the free wall of the LV. The impact of CRT is influenced by the presence of LV activation delay.<sup>7</sup> Secondly, the correction of atrioventricular dyssynchrony by correcting the atrioventricular conduction time reduces the filling diastolic pressure of the LV and the amount of mitral re-

gurgitation.<sup>8</sup> Thirdly, the resynchronization of papillary muscle contraction reduces systolic mitral regurgitation.<sup>9</sup> Finally, long-term antiarrhythmic control could protect against ventricular tachyarrhythmias.<sup>6</sup> By applying all these benefits of CRT pacing, an inverted remodeling and amelioration of ejection fraction can be observed in patients who respond to therapy.

SSc is a systemic disease that involves not only the skin but also the cardiovascular system. The cardiac manifestations of SSc include myocardial fibrosis due to inflammatory, fibrotic, and vasculopathic changes at the level of the myocardium.<sup>10</sup> Fibrosis generally affects both ventricles and atria, probably due to microvascular dysfunction and perivascular inflammation.<sup>11,12</sup> These changes can be observed in up to 80% of patients with SSc. The functional manifestations of LV myocardial fibrosis, such as LV diastolic dysfunction and contractile abnormalities, can be observed on 2D echocardiography.<sup>13</sup> Severe systolic dysfunction with a reduced ejection fraction is rarely seen compared with impaired cardiac relaxation.<sup>14</sup> Structural modifications in the epicardial coronary arteries may initiate intermittent myocardial hypoxia and consequently can accelerate the progression and extension of myocardial fibrosis.<sup>15</sup> The progression of fibrosis can also affect the conduction system of the heart, the presence of different types of atrioventricular block or ventricular ectopic beats being related to a negative prognosis.<sup>16</sup>

Elevated NT-proBNP levels and reduced LVEF are the strongest predictors of mortality in patients with SSc.<sup>17</sup> LV dysfunction along with perfusion defects on SPECT were associated with the development of aggravated HF and increased mortality rate.<sup>18</sup> CRT may be crucial in selected patients with SSc (about 5% of which present HF with reduced ejection fraction [HFREF]) because of its benefits on electric and mechanical resynchronization.

This case emphasizes the importance of CRT implantation in patients suffering from SSc, even if the operator can expect more difficulties during the procedure. Technical issues, such as difficult cannulation of the CS or vein occlusion, may suggest the use of alternative tools from the invasive cardiology toolkit to overcome these difficulties. From an anatomical point of view, the persistence of Thebesian valve or the presence of a slightly more distally located Marshall valve can make it more difficult to properly position the LV lead.<sup>19</sup> In most cases, CCTA or an angiogram performed in the venous phase of coronary angiography can enable a comprehensive assessment and preparation. The presence of valves in the CS vein often requires balloon venoplasty. In this case, we used a balloon typically employed in the arterial system. Stenting

can also be used to position the LV lead and can provide long-term assurance that the lead remains in the proper position, which is critical for resynchronization. However, if extraction were to be necessary, stenting can make the procedure more challenging.<sup>20</sup> Furthermore, it is important to emphasize that the use of arterial devices at the venous level always requires increased attention and caution, as the risk of perforation and dissection is higher due to its different structure and thinner wall.

This case underlines the importance of anatomical preparation with appropriate imaging investigations for proper planning of the interventional approach. There are major benefits of CRT implantation in selected patients with SSc and HF<sub>rEF</sub>. However, the operator should always be prepared for anatomical difficulties based on the character of the SSc. Our case demonstrates that even in critical cases with decompensated HF and associated SSc, which make the CRT implantation more demanding, this therapy may significantly improve the clinical outcome.

## CONCLUSION

In patients with decompensated HF and SSc, CRT implantation may improve prognosis and quality of life. In rare cases, CS occlusion makes implantation of the LV pacing electrode extremely difficult; these cases can be managed using percutaneous balloon angioplasty.

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## CONFLICT OF INTEREST

Nothing to declare.

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