Is the epicardial left ventricular lead implantation an alternative approach to percutaneous attempt in patients with Steinert disease? A case report

Andrea Antonio Papa1, 2, Anna Rago1, Roberta Petillo2, Paola D’Ambrosio2, Marianna Scutifero2, Marisa De Feo3, Ciro Maiello4 and Alberto Palladino2

1 Department of Cardiothoracic Sciences, Chair of Cardiology, Monaldi Hospital, University of Campania "L. Vanvitelli"; 2 Cardiomyology and Medical Genetics, University Hospital of Campania “L. Vanvitelli”; 3 Department of Cardiothoracic Sciences, Unit of Cardiac Surgery, Monaldi Hospital; 4 Transplant Surgery Unit, AORN Ospedali dei Colli, Monaldi Hospital, Naples, Italy

Steinert’s disease or Myotonic Dystrophy type 1 (DM1) is an autosomal dominant multisystemic disorder characterized by myotonia, muscle and facial weakness, cataracts, cognitive, endocrine and gastrointestinal involvement, and cardiac conduction abnormalities. Although mild myocardial dysfunction may be detected in this syndrome with age, overt myocardial dysfunction with heart failure is not frequent. Cardiac resynchronization therapy is an effective treatment to improve morbidity and reduce mortality in patients with DM1 showing intra-ventricular conduction delay and/or congestive heart failure. We report the case of a patient with Steinert disease showing an early onset ventricular dysfunction due to chronic right ventricular apical pacing, in which an epicardial left ventricular lead implantation was performed following the failure of the percutaneous attempt. As no relief in symptoms of heart failure, nor an improvement of left ventricular ejection fraction and reverse remodelling was observed six months later, the patient was addressed to the heart transplantation.

Key words: cardiac resynchronization, epicardial left ventricular implantation, Steinert disease

Introduction

Myotonic dystrophy type 1 (DM1) or Steinert’s disease, is the most common muscular dystrophy in adult life with an estimated prevalence of 1/8000. Cardiac involvement, including conduction abnormalities with arrhythmias and conduction disorders, contributes significantly to the morbidity and mortality of the disease. It is recorded in about 80% of cases, and may precede the involvement of skeletal muscles (1-3). The characteristic impairment of His-Purkinje system is the most common cardiac abnormality. Mild ventricular dysfunction has also been reported associated with conduction disorders, but severe ventricular systolic dysfunction is not frequent and usually occurs late in the course of the disease as the final stage of cardiomyopathy (1). Cardiac resynchronization therapy (CRT) is able to restore physiological pattern of ventricular depolarization, resulting in reduction of mitral regurgitation and improvement of left ventricular (LV) systolic function (4-6). CRT has demonstrated reduction in morbidity and mortality in patients with severe refractory heart failure (HF) and intraventricular conduction delay (4-6). The technique of choice for left ventricular pacing in ventricular resynchronization is the insertion of a lead through the coronary sinus, into the posterolateral vein. The epicardial placement of ventricular leads is considered at present, a salvage technique for patients in whom the percutaneous procedure fails (7).

We report the case of a patient with Steinert disease showing an early onset ventricular dysfunction caused by chronic right ventricular apical pacing, in which an epicardial left ventricular lead implantation was performed following the failure of the standard percutaneous attempt. As no relief in symptoms of heart failure, nor
an improvement of left ventricular ejection fraction and reverse remodelling was observed six month later, the patient has been addressed to the heart transplantation.

Case report

A 43-year-old man – affected by Steinert disease and regularly followed at Cardiomyology and Medical Genetics Service since the time of his diagnosis (2003) – was hospitalised for an exacerbation of signs and symptoms of congestive heart failure [fatigue, muscle weakness, dyspnea, orthopnea, edema and palpitations, New York Heart Association (NYHA) class III]. His blood pressure (BP) was 100/60 mmHg and heart rate (HR) 60/bpm. Crackles at the basal field of lungs and pretibial edema were detected. Chest X-ray revealed cardiac dilation and pulmonary congestion.

The diagnosis of DM1, at first based on the family history (one affected brother) and clinical features (myotonic phenomenon, mild distal skeletal muscle dysfunction, cataract, gastrointestinal disturbances, endocrine deficiency), was subsequently confirmed by molecular testing, that showed a pathological expansion of CTG triplets (E1 class). In 2005, a bicameral pacemaker (PM) was implanted because evidence of first degree (PR interval ≥ 255ms) plus second-degree type2 atrio-ventricular block (8-13), and concomitant paroxysmal atrial flutter (AF) episodes. The implant was made according to the current guidelines (14) and was followed by an improvement of symptoms and quality of life. To be noted that atrial arrhythmias are not rare in this population (15-17).

In 2013, the PM – according to the current guidelines (18) – was uploaded to a cardioverter defibrillator (ICD) due to the finding of not sustained ventricular tachycardia (NSVT) in pacemaker stored electrograms to prevent the high risk of sudden cardiac death, frequently observed in these patients as in others muscular dystrophies (19). The ICD was placed in the right position, because of the occlusion of left subclavian vein (20-22).

In 2016 during a routine clinical and instrumental follow-up, signs of congestive heart failure (CHF) were detected. The ECG showed a sinus rhythm and a wide QRS interval (165 ms) due to constant right ventricular apical pacing (Fig. 1). Transthoracic echocardiography showed dilation of the heart (left ventricular end-diastolic diameter – LVEDD – was 7.4 cm), left systolic dysfunction and overt intra- and inter-ventricular asynchrony. The ejection fraction (EF), calculated by the Simpson and Teichholz method, was 25% (Fig. 2).

The interrogation of ICD revealed absence of intrinsic spontaneous ventricular rhythm, not sustained paroxysmal episodes of atrial flutter/fibrillation and ventricular tachycardias and no episodes of malignant sustained ventricular arrhythmias requiring device intervention. According to the current guidelines (23), his medical therapy was adjusted and included aggressive loop diuretic therapy, β-blockers, spironolactone and ACE inhibitors. In order to rule out an ischemic aetiology of dilated cardiomyopathy and consequent heart failure, a diagnostic coronary angiography was performed showing normal coronary arteries. Despite the aggressive medical therapy the patient experienced two episodes of acute heart failure over one year period, posing the indication for cardiac resynchronization therapy by a biventricular ICD-CRT (24). Before the intervention, a right subclavian venogram was performed which revealed a long segment of occlusion; any attempt to recanalise the right subclavian vein percutaneously failed. Venous stenosis or occlusion due to thrombosis/fibrosis resulting from the presence of the lead is a frequent side-effect in patients implanted with devices. In these cases, an epicardial approach is planned.

Technical procedure

The procedure was performed after written informed consent. Left antero-lateral thoracotomy was performed
along the fourth intercostal space under general anesthesia. The patient was placed in a 45° rotation to the right side. A 3- to 4- cm long minithoracotomy was performed through the fourth intercostal space between the anterior and mid-axillary line.

The pericardium was opened longitudinally anterior to the phrenic nerve and suspended with traction sutures to better expose the lateral wall. The epicardial lead (bipolar) was fixed at the anterolateral wall of LV. Electrical parameters were measured to verify the correct positioning of the new leads. Once a site with satisfactory pacing threshold was identified (impedance > 200 Ω and < 2000 Ω, sensing > 5 mV and pacing threshold measured at 0.5 ms < 2.0 V), the lead was sewn with 5/0 polypropylene sutures. The connector of the lead was tunelled to the ICD-CRT device pocket in the right pre-pectoral region. The previous endocardial right ventricular defibrillation lead was connected to the ICD-CRT generator (Fig. 3). The patient was extubated in the operating room and observed in the cardiac surgery recovery unit for 24 hours.

**Patient’s follow-up**

The post-operative follow up included the assessment of NYHA functional class, ECG with determination of QRS duration and echocardiography. Left ventricular ejection fraction, left ventricular end-diastolic dimension and severity of mitral regurgitation (MR) values were collected to analyse the effect of CRT via epicardial LV lead placement on reverse ventricular remodelling. One month later an optimization of the atrio-ventricular and inter-ventricular intervals during cardiac resynchronization was performed by both ECG and echocardiogram.

At six-months follow-up, no relief of symptoms was reported by the patient. In that occasion ECG revealed paced biventricular rhythm with a still wide QRS interval (150 ms, Fig. 4), though of reduced size compared to the previous one, and no changes in the repolarization dispersion time. Despite an adequate biventricular pacing the patient remains in NYHA class-III and experienced a further episode of acute heart failure requiring hospitalization. The echocardiogram didn’t show an improvement of EF and LV stroke volume (Fig. 5). The ICD analysis showed no significant modification of the electrical parameters, paroxysmal atipical atrial flutter/fibrillation and 98% biventricular pacing rhythm. The patient experienced one episode of slow sustained monomorphic ventricular tachycardia (140 bpm), recognized in monitoring zone of the device, which required external electrical cardioversion.

**Discussion**

Conduction abnormalities are the most frequent finding of cardiac involvement in patients with DM1 and minor conduction defects can be present in early stages of the disease (1, 2, 25-27). More severe conduction defects may be cause of shortness of breath, dizziness, faint-
ing, syncope, and even of sudden death. Left ventricular dilatation with overt systolic dysfunction is not frequent; however when present they may be more prominent than the muscle complaint. Cardiac symptoms generally occur later compared to the skeletal muscle weakness, but sometimes they may be the initial manifestation of the disease (1, 2, 25). In patient here reported, the early onset of heart failure could be related to the electromechanical delay caused by both intra- and inter-ventricular asynchrony due to chronic right apical pacing; the latter leads to regional structural changes causing a uncoordinated heart contraction that in turn accelerates the progression of the heart failure (28). Beside advances in the optimal medical treatment, strategies for medically refractory symptomatic advanced heart failure have emerged, including cardiac resynchronization therapy. Patients with NYHA class III or IV, with EF 35% or less, sinus rhythm with a QRS duration ≥ 130 ms and left bundle branch block (LBBB) or a QRS duration ≥ 150 ms irrespective of the QRS morphology, are eligible to receive a cardiac resynchronization therapy, according to the current guidelines (23). Basing on the progression of LV dysfunction, AV conduction disturbances and the frequent occurrence of ventricular tachyarrhythmia, Said et al. (29) hypothesized a role for biventricular ICD in patients with DM1 who need a permanent pacemaker implantation. Two previous papers (7, 30) reported an improvement in symptoms of heart failure, LVEF and reverse remodelling in one patient with DM1 showing an early onset ventricular dysfunction secondary to a complete LBBB by this approach. However, a clear consensus about biventricular pacing or the usage of ICD does not exist for this kind of patients.

The “standard of care” of left lead implantation for CRT still remains the less invasive transvenous approach (31). However, several issues may result in failed transvenous implantation of the LV lead such as anatomical limitations due to occlusion of the subclavian vein or the superior vena cava, or an abnormal anatomy of the coronary sinus. Furthermore, lead-related issues such as lead instability with repeated dislodgement, phrenic nerve stimulation despite electrical or physical optimization, or systemic conditions such as endocarditis may contribute to failed transvenous LV lead implantation (31). In these cases, the surgical placement of an epicardial LV lead is required with satisfactory long-term results (32).

Conclusions

The case here reported is the first patient with DM1 in which an epicardial left ventricular lead implantation was used for cardiac resynchronization therapy after failure of percutaneous attempt. At six-months follow-up, based on this experience, the epicardial CRT did not induce either symptom relief, nor improvement of the ejection fraction or reduction of the arrhythmic risk. A possible explanation of the heart failure in this patient may be the prolonged apical pacing; further studies are in progress to determine the consequences of long-term constant apical pacing in patients affected by Myotonic Dystrophy type 1.

References


