Journal of Zhejiang University-SCIENCE B (Biomedicine & Biotechnology)
ISSN 1673-1581 (Print); ISSN 1862-1783 (Online)
www.zju.edu.cn/jzus; www.springerlink.com
E-mail: jzus@zju.edu.cn

Letters:

Application of CRT-D in a Marfan syndrome patient with chronic heart failure accompanied by ventricular tachycardia and ventricular fibrillation

Hai-ying XU[†1], Yi-zhou XU[1], Feng LING[1], Zhong YU[1], Jun YANG[1], Xu DUAN[1], Bei WANG[2], Jin-yu HUANG[†‡1]

(*[1]Department of Cardiology, Hangzhou First People's Hospital, Hangzhou 310006, China*)

(*[2]Department of Cardiac Ultrasound, Hangzhou First People's Hospital, Hangzhou 310006, China*)

[†]E-mail: 1339220837@qq.com; hjyuo@163.com

Received July 3, 2013; Revision accepted July 20, 2013
Crosschecked July 3, 2013

doi:10.1631/jzus.BQICC710
Document code: A **CLC number:** R459.9

Marfan syndrome is a systemic connective tissue disease that could affect the cardiovascular system and eventually lead to heart enlargement and heart failure with high mortality, mainly due to progressive heart failure and/or sudden cardiac death caused by malignant arrhythmia. Here we report that a patient received a cardiac resynchronization therapy-defibrillator (CRT-D) with a pre-monitor function for heart failure and experienced obvious improvements in his cardiac function. Postoperative follow-up showed that the patient had reduced morbidity and hospitalization for heart failure, and also experienced improved quality of life.

A 69-year-old male patient was admitted six months ago due to coughing, and expectoration with a fever for four days. The patient was diagnosed with Marfan syndrome and accepted ascending aorta and had an aortic valve replacement surgery 14 years ago. With a 10-year history of hypertension and a cerebral infarction 5 years ago, he had been suffering repeated symptoms of heart failure during the past year, and was hospitalized after diagnosis of lung infection, worsening heart failure and atrial fibrillation. On the night of admission he went through sudden cardiac arrest of the ventricular fibrillation, and survived after immediate defibrillation and resuscitation procedures, with heart rhythm restored through atrial fibrillation.

Physical examination revealed jugular vein filling, and diminished vesicular sounds on both sides of the chest. The heart rate was irregular at 70 beats/min (bpm). II/6 grade systolic murmur (SM) at mitral area and III/6 grade SM at aortic valve area were also heard. The heart was enlarged and no edema was found in either legs.

A chest X-ray showed right lower lung infection and pleural effusion, with a cardiothoracic ratio of 0.65; B ultrasound showed a bilateral pleural effusion (median volume); echocardiography showed a left ventricular enlargement (left atrium 5.2 cm, left ventricular 6.4 cm) and a left ventricular dysfunction (ejection fraction (EF) 0.11); sequential electrocardiographs (ECGs) showed sinus rhythm with atrial fibrillation, complete left bundle branch block (CLBBB), and a left ventricular hypertrophy with strain.

On the basis of inotropes, digoxin, lotensin, diuretics, amiodarone, warfarin, and a small dose of carvedilol as well as antibiotics, he received an implant INSYNC SENTRYTM7298 CRT-D (Bristow *et al.*, 2004). The parameters are listed in Table 1. Due to the patient's severe condition, a defibrillation threshold test (DFT) was not carried out during the procedure. The settings of the tachycardia diagnostic criteria were: ventricular tachycardia (VT), 130–150 bpm, monitor; fast ventricular tachycardia (FVT), 150–180 bpm, antitachycardia pacing (ATP); ventricular fibrillation (VF), >182 bpm, shock.

‡ Corresponding author

Table 1 Parameters for each lead

Lead	Threshold (V)	Impedance (Ω)	Amplitude (mV)
Left ventricle	0.5	769	17.9
Right ventricle	0.5	1280	14.2
Right atria	Not tested (AF)	560	2.6

The patient still felt chest tightness and shortness of breath two days after the procedure, and complained of shock in the morning of the third day. The programmer showed 5 VF events, which led to 6 shock therapies (two shocks for one of the events), and all events were completed successfully. The patient continued to receive optimized drug therapy. Thereafter, an ECG showed atrial fibrillation and biventricular pacing, with QRS wave duration of 150 ms (Chung *et al*., 2008). His sinus rhythm was restored after repeated defibrillation therapies due to recurrent ventricular tachycardia ventricular fibrillation. In the beginning, the sinus rhythm could only be sustained for quite short periods of time, but with administration of amiodarone and carvedilol, sustained atrial fibrillation gradually converted into paroxysmal atrial fibrillation, and finally converted into sustained sinus rhythm. Twenty days after the implantation of CRT-D, the patient felt well.

One month after the implantation, parameter optimization was carried out guided by tissue Doppler imaging (TDI) (Barsheshet *et al*., 2011). The left ventricular filling was evaluated by mitral time-velocity integral (TVI) during the diastolic phase (Dohi *et al*., 2005), and mitral regurgitation was evaluated by TVI of the mitral regurgitation. The optimal atrioventricular (AV) interval was identified through an increased left ventricular filling and reduced mitral regurgitation, i.e., 100 ms for sensed atrioventricular (SAV) and 130 ms for paced atrioventricular (PAV). The time to peak myocardial systolic velocity at septum and lateral left ventricular wall was recorded from the TDI (Penicka *et al*., 2004), and the difference between them was calculated to evaluate the desynchrony; the TVI of the aortic outflow was recorded as an indicator of left ventricular output (Mele *et al*., 2006). The optimal left ventricular to right ventricular (VV) interval was defined as that which could best restore inter- and intra-ventricular synchrony. The test showed that when the VV interval (Baker *et al*., 2007) was set to 32 ms, the difference between the time to peak myocardial systolic velocity at septum and the lateral left ventricular wall decreased to 63 ms from 176 ms before optimization (Fig. 1).

Echocardiography parameters six months after implantation are shown in Table 2. A built-in compass of CRT-D showed that the heart rate gradually declined and stabilized at 60 to 70 bpm with increases in the patient's daily activities. The pre-monitor function of the heart failure in six months after the operation showed that transthoracic impedance gradually increased and stabilized, indicating significant improvement of the pulmonary edema.

Table 2 Echocardiographic parameters before and after implantation

Group	LAD (mm)	LVEDD (mm)	LVEF	NYHA class	QRS duration (s)
Before	51.69	63.72	0.11	4	0.15
After	39.20	64.20	0.45	2–3	0.15

LAD: left atrial diameter; LVEDD: left ventricular end-diastolic diameter; LVEF: left ventricular ejection fraction; NYHA: New York Heart Association

The patient was suffering a severe condition, which led to repeated malignant arrhythmia and defibrillation. Thus we proposed that timely implantation of CRT-D would bring more benefit for the patient despite a higher risk with these procedures.

The efficacy of implantable cardioverter defibrillator (ICD) for primary and secondary prevention of sudden cardiac death (SCD) has been proven in a large number of randomized controlled trials. In some trials, DFT was carried out to ensure that ICD or CRT-D could appropriately cease ventricular arrhythmias. Nevertheless, Russo *et al*. (2005) suggested that a simple application of a defibrillator with high output (≥35 J) in 48% of the study patients (3% of the entire installation ICD crowd) could not get the appropriate DFT, and 6% of ICD recipients had excessive DFT (safety margin <10 J). This patient did not test intraoperative DFT due to a severely impaired cardiac function, but the defibrillation was successful with the highest energy to prevent SCD. There is no unified standard to decide which patient population should undergo a scheduled DFT, but it has been suggested that patients with a left ventricular diastolic diameter ≥80 mm or with severely impaired heart function should suspend the DFT to ensure their safety during the procedure.

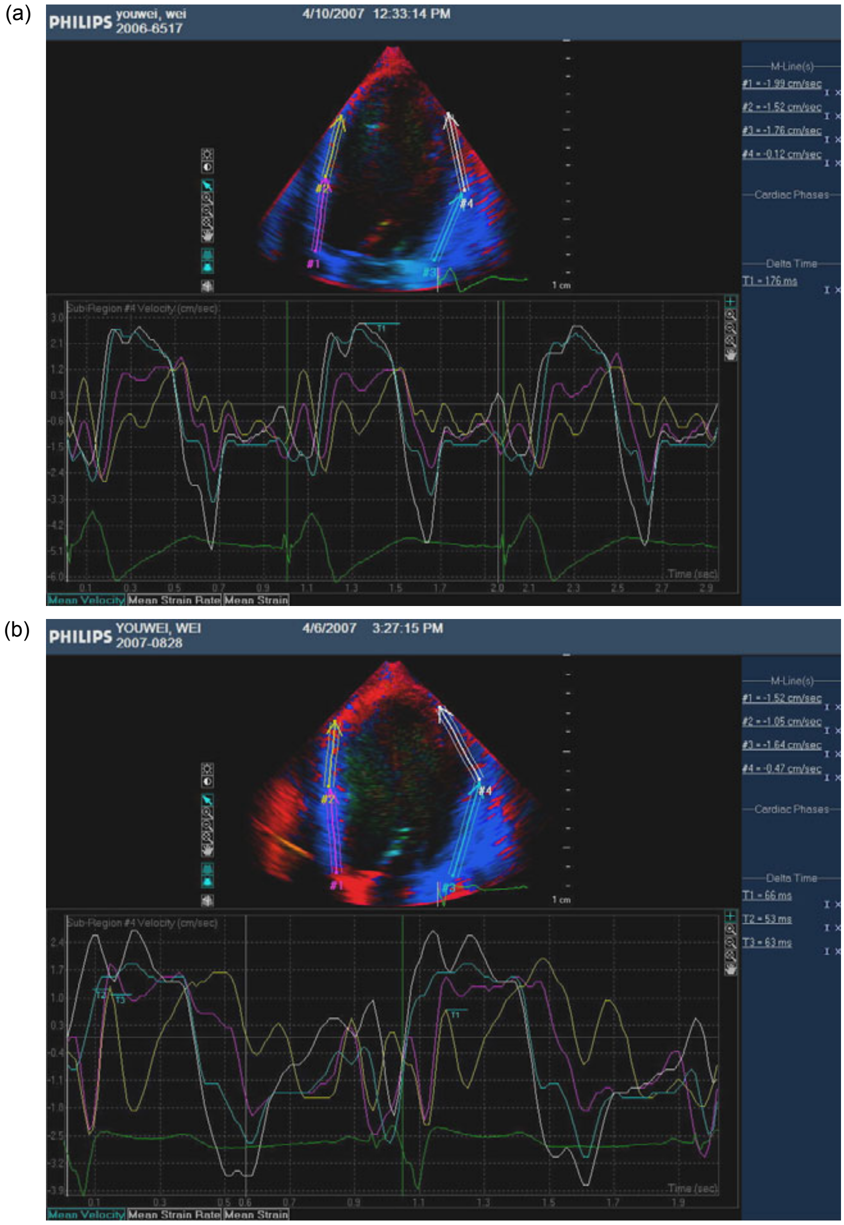

Fig. 1 TDI before (a) and after (b) optimization
(a) Difference between time to peak myocardial systolic velocity at septum and lateral left ventricular wall was 176 ms; (b) When the VV interval was set to 32 ms, the difference between the time to peak myocardial systolic velocity at septum and the lateral left ventricular wall was shortened to 63 ms

The patient was diagnosed as atrial fibrillation with obviously enlarged left atria, and was generally considered not necessary or feasible for cardioversion. For CRT candidates with permanent atrial fibrillation, it is generally accepted that atrioventricular node ablation is a good way to ensure dual-chamber pacing. This patient in our report initially suffered recurrent ventricular tachycardia, and sinus rhythm restored after defibrillation therapy could only be sustained for a quite short period of time. With administration of amiodarone and carvedilol, and improvement in cardiac function, sustained atrial fibrillation gradually converted into paroxysmal atrial fibrillation, and finally converted into sustained sinus rhythm 20 d after the implantation of CRT-D. This prompted us to take a review of the patients with heart failure and atrial fibrillation that were believed to be appropriate to receive cardioversion, and to evaluate the probability and necessity of these patients to go through medical and/or electrical cardioversion after CRT. Especially, if early restoration of sinus rhythm in these patients could help to maintain the atrioventricular synchrony, it would be more valuable for long-term prognosis improvement.

This patient received CRT-D with pre-monitor function of heart failure, with the unique OptiVol™ technology that could accurately monitor the status of pulmonary edema during heart failure. For this patient, we found that the transthoracic impedance gradually reached steady status with improvement in cardiac function during follow-up. We believe that this would help patients and doctors to take timely measures to reduce morbidity and hospitalization for heart failure, and improve quality of life (Suffoletto *et al*., 2006), and finally improve prognosis.

Echocardiography is playing an increasingly important role in the study of patients with clinical and echocardiographic criteria for cardiac resynchronization therapy. The case presented highlights the importance of intraventricular dyssynchrony, identified by TDI. In this case through VV optimization, the difference between the time to peak myocardial systolic velocity at the septum and lateral left ventricular wall was shortened to 63 ms, which showed that the intraventricular dyssynchrony was improved and that the haemodynamics got better.

From what we discussed above, we can draw the conclusion that CRT-D made sense for this patient. The patient after the synchronization, showed ventricular hemodynamic improvement resulting in improvement in the cardiac function. VF was effectively controlled to improve the arrhythmia, and the cardiac function was improved thereby. A strong adaptation of preoperative choice is also an important aspect for the patient.

Compliance with ethics guidelines

Hai-ying XU, Yi-zhou XU, Feng LING, Zhong YU, Jun YANG, Xu DUAN, Bei WANG, and Jin-yu HUANG declare that they have no conflict of interest.

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000(5). Informed consent was obtained from all patients for being included in the study.

References

Baker, J.H., McKenzie, J., Beau, S., Greer, G.S., Porterfield, J., Fedor, M., Greenberg, S., Daoud, E.G., Corbisiero, R., Bailey, J.R., *et al*., 2007. Acute evaluation of programmer-guided AV/PV and VV delay optimization comparing an IEGM method and echocardiogram for cardiac resynchronization therapy in heart failure patients and dual-chamber ICD implants. *J. Cardiovasc. Electrophysiol.*, **18**(2):185-191. [doi:10.1111/j.1540-8167.2006.00671.x]

Barsheshet, A., Goldenberg, I., Moss, A.J., Eldar, M., Huang, D.T., McNitt, S., Klein, H.U., Hall, W.J., Brown, M.W., Goldberger, J.J., *et al*., 2011. Response to preventive cardiac resynchronization therapy in patients with ischaemic and nonischaemic cardiomyopathy in MADIT-CRT. *Eur. Heart J.*, **32**(13):1622-1630. [doi:10.1093/eurheartj/ehq407]

Bristow, M.R., Saxon, L.A., Boehmer, J., Krueger, S., Kass, D.A., de Marco, T., Carson, P., DiCarlo, L., DeMets, D., White, B.G., *et al*., 2004. Cardiac-resynchronization therapy with or without an implantable defibrillator in advanced chronic heart failure. *N. Engl. J. Med.*, **350**(21):2140-2150. [doi:10.1056/NEJMoa032423]

Chung, E.S., Leon, A.R., Tavazzi, L., Sun, J.P., Nihoyannopoulos, P., Merlino, J., Abraham, W.T., Ghio, S., Leclercq, C., Bax, J.J., *et al*., 2008. Results of the predictors of response to CRT (PROSPECT) trial. *Circulation*, **117**(20): 2608-2616. [doi:10.1161/CIRCULATIONAHA.107.743120]

Dohi, K., Suffoletto, M., Ganz, L., Zenati, M., Gorcsan, J., 2005. Utility of echocardiographic tissue synchronization imaging to redirect left ventricular lead placement for improved cardiac resynchronization therapy. *Pacing Clin. Electrophysiol.*, **28**(5):461-465. [doi:10.1111/j.1540-8159.2005.40056.x]

Mele, D., Pasanisi, G., Capasso, F., de Simone, A., Morales, M.A., Poggio, D., Capucci, A., Tabacchi, G., Sallusti, L., Ferrari, R., 2006. Left intraventricular myocardial deformation dyssynchrony identifies responders to cardiac resynchronization therapy in patients with heart failure. *Eur. Heart J.*, **27**(9):1070-1078. [doi:10.1093/eurheartj/ehi814]

Penicka, M., Bartunek, J., de Bruyne, B., Vanderheyden, M., Goethals, M., de Zutter, M., Brugada, P., Geelen, P., 2004. Improvement of left ventricular function after cardiac resynchronization therapy is predicted by tissue Doppler imaging echocardiography. *Circulation*, **109**(8):978-983. [doi:10.1161/01.CIR.0000116765.43251.D7]

Russo, A.M., Sauer, W., Gerstenfeld, E.P., Hsia, H.H., Lin, D., Cooper, J.M., Dixit, S., Verdino, R.J., Nayak, H.M., Callans, D.J., *et al*., 2005. Defibrillation threshold testing: is it really necessary at the time of implantable cardioverter-defibrillator insertion? *Heart Rhythm.*, **2**(5): 456-461. [doi:10.1016/j.hrthm.2005.01.015]

Suffoletto, M.S., Dohi, K., Cannesson, M., Saba, S., Gorcsan, J., 2006. Novel speckle-tracking radial strain from routine black-and-white echocardiographic images to quantify dyssynchrony and predict response to cardiac resynchronization therapy. *Circulation*, **113**(7):960-968. [doi:10.1161/CIRCULATIONAHA.105.571455]