Use of Biventricular Pacing in Arrhythmogenic Right Ventricular Cardiomyopathy with Disarticulated Right Ventricle

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Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterised by fibrofatty replacement of cardiomyocytes and is associated with structural and functional abnormalities of the right ventricle (RV). These may include regional wall motion abnormalities, ventricular aneurysms, ventricular dilation and dysfunction leading either to right sided heart failure, or may also affect the left ventricle (LV) leading to biventricular heart failure. Electrocardiographic depolarisation/repolarisation abnormalities and ventricular tachyarrhythmias are also typically present. In the early stages of the disease structural changes are typically localised to the right ventricular outflow tract, inflow tract or apex of the RV known as the “triangle of dysplasia” (Marcus et al., 2010).

Clinical presentation usually includes palpitations, chest pain, syncope, ventricular tachycardia (VT) with left bundle branch block morphology and sudden death. ARVC has a reported overall prevalence of approximately 1 in 1000 to 1 in 5000 (Hamilton, 2009) and is a significant cause of sudden cardiac death in young people and athletes.

ARVC is a complex disease and is difficult to diagnose. An international task force established a set of diagnostic criteria in 1994 (McKenna et al., 994) which was later reviewed and modified in 2010 (Marcus et al., 2010) to include new diagnostic modalities and technology and to improve diagnostic sensitivity. The criteria involve a scoring system looking at RV morphological and functional abnormalities, electrocardiogram (ECG) features, histopathological findings, ventricular arrhythmias and family history.

Ablating a single site of ventricular tachycardia in ARVC patients is unlikely to eliminate the arrhythmia as the entire right ventricle is potentially arrhythmogenic therefore right ventricular disarticulation can be used to prevent multifocal VT arising from the right ventricular free wall (RVFW). This procedure was first described by Guiraudon et al. in 1983 and involves surgically dissecting the RVFW from its LV attachments. The RVFW is then sutured back in place and the fibrous scar formed electrically isolates the RVFW confining any VT to that chamber alone.

Case Presentation

A 41-year-old male with a history of ARVC and subsequent RV disarticulation procedure to treat intractable ventricular arrhythmias (performed when the patient was aged 23) attended cardiac outpatients for routine follow-up. The patient also had a history of recurrent atrial arrhythmias of which he was symptomatic and had

required repetitive ablation procedures, one of which had been complicated by complete heart block requiring insertion of permanent dual chamber pacemaker.

He had initially felt symptomatic benefit following his last ablation procedure which was a redo-atrial flutter ablation, but had since felt a gradual decline and was experiencing intermittent fast palpitations associated with breathlessness, light-headedness and chest/throat tightness. He reported no syncopal episodes but had noticed some ankle swelling for which he had taken a stat dose of Furosemide which had helped significantly.

Pacing check revealed further episodes of atrial flutter and timings were consistent with his symptoms. Echocardiogram showed marked right heart dilatation with very severe RV systolic dysfunction and free tricuspid regurgitation due to annular dilatation. Marked paradoxical septal motion was present and the inferior vena cava (IVC) was dilated with less than 50% collapse and systolic flow reversal seen in the dilated hepatic veins.

The recurrence of his symptomatic atrial arrhythmias despite several previous ablation attempts and new symptoms associated with right sided heart failure led to discussion at multidisciplinary team meeting. It was felt that his RV was virtually static with the tricuspid valve (TV) remaining open throughout the cardiac cycle and the RV therefore acting similarly to a conduit from the right atrium (RA) to pulmonary arteries. Consequently, advice was gathered regarding any possible conventional congenital approaches that might benefit his cardiac function. Cavopulmonary anastomosis was considered in order to offload the right heart and TV replacement was also considered. However, it was felt these options were unlikely to be of any benefit and were extremely hazardous from a surgical risk.

The patient was still highly symptomatic of atrial arrhythmias and had been programmed to DDI mode on his pacemaker to avoid tracking of these arrhythmias, however it was unclear whether some of his lethargy was being caused by lack of atrioventricular (AV) synchrony when in sinus rhythm due to this non-tracking mode.

The patient went back to the electrophysiology (EP) lab to assess and attempt to ablate his atrial arrhythmias. The patient was in atrial flutter at the start of the procedure. Carto mapping system demonstrated large areas of scar appearing to relate to both surgical incision and dilatation. The flutter circuit which demonstrated early signals in the lateral RA was treated with radiofrequency ablation which joined the narrowest scar and the IVC. Re-induction produced other atrial arrhythmias and it appeared these would be difficult to manage completely. During the EP study, the RVFW was assessed and demonstrated that it could be independently captured.

The patient had short runs of atrial tachycardia overnight and was commenced on 50mg Flecainide twice daily the next morning which the patient failed to tolerate and quickly stopped taking. After a week, the patient was still very lethargic and the rate response settings on his pacemaker were optimised. The mode switch rate on his pacemaker could not be programmed lower than 130bpm, however his atrial tachycardia had a rate of less than 130bpm therefore the device was programmed DDIR to prevent tracking of the arrhythmia. He was started on Propafenone.

Another week later the patient was still feeling lethargic and experiencing the neck tightness sensation, but heart rate histograms from the pacemaker suggested his atrial rate was now controlled and the pacemaker was programmed back to DDD mode in the hope that restoration of AV synchrony would improve his symptoms.

The patient got in touch with the department to report that he was still symptomatic and following discussion with the consultant the patient was admitted for elective upgrade to cardiac resynchronisation therapy pacemaker (CRT-P). A second RV lead was implanted in the RVFW and the device upgraded from a dual chamber pacemaker to a CRT-P device (Figures 1-3). Please see Table 1 for the programming of the device after echocardiography optimisation which was performed to resynchronise the disarticulated RVFW with the RV apex.

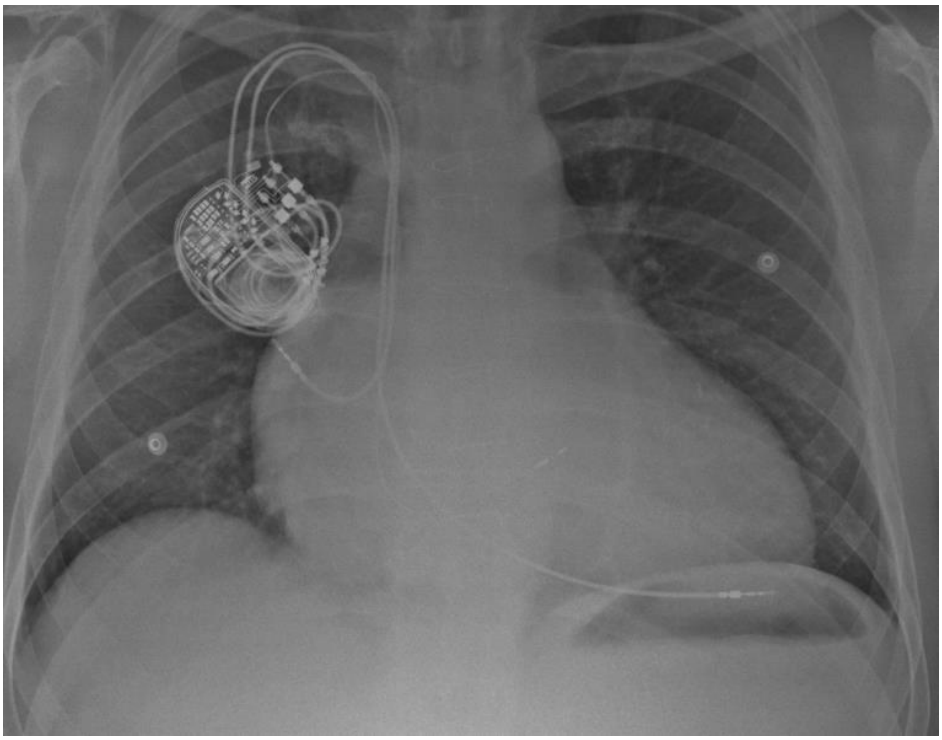

Figure 1: X-ray demonstrating right sided CRT-P implant with two ventricular leads in the right ventricle, one in the RV apex and one in the RVFW.

Table 1: CRT-P programming after optimisation using echocardiography to synchronise right ventricular contraction.

Mode	DDD
Lower rate	60 bpm
V-V pacing delay	LV-RV 60ms (lead in the disarticulated RVFW in LV port)

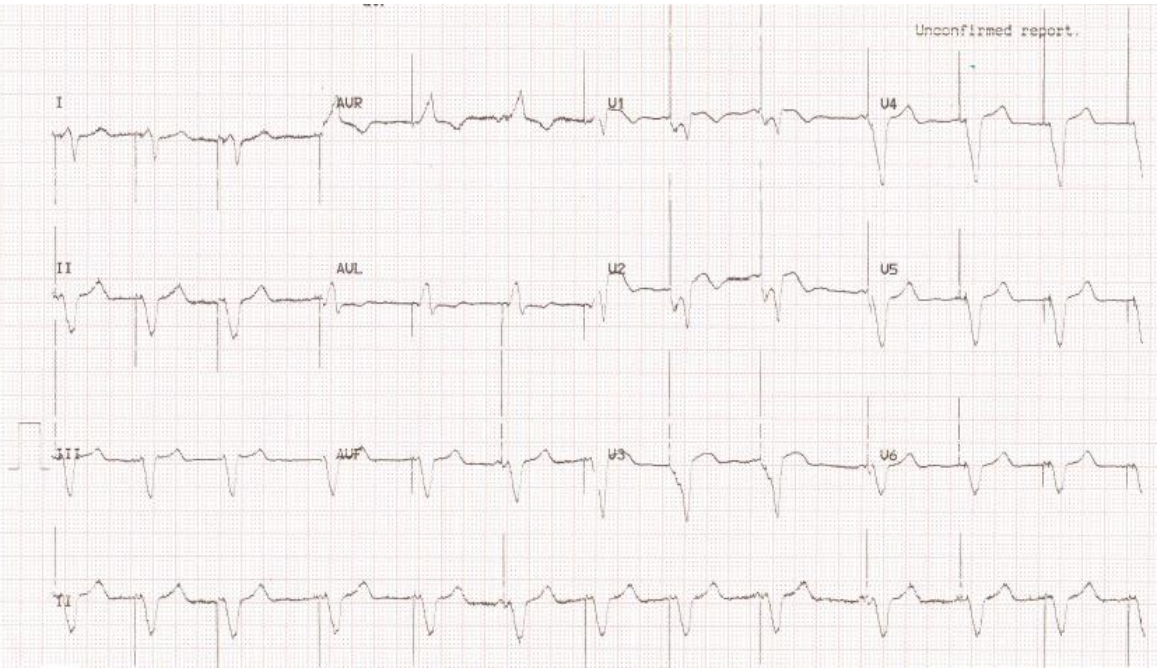

Figure 2: ECG showing multisite RV pacing in the RV apex and RV free wall.

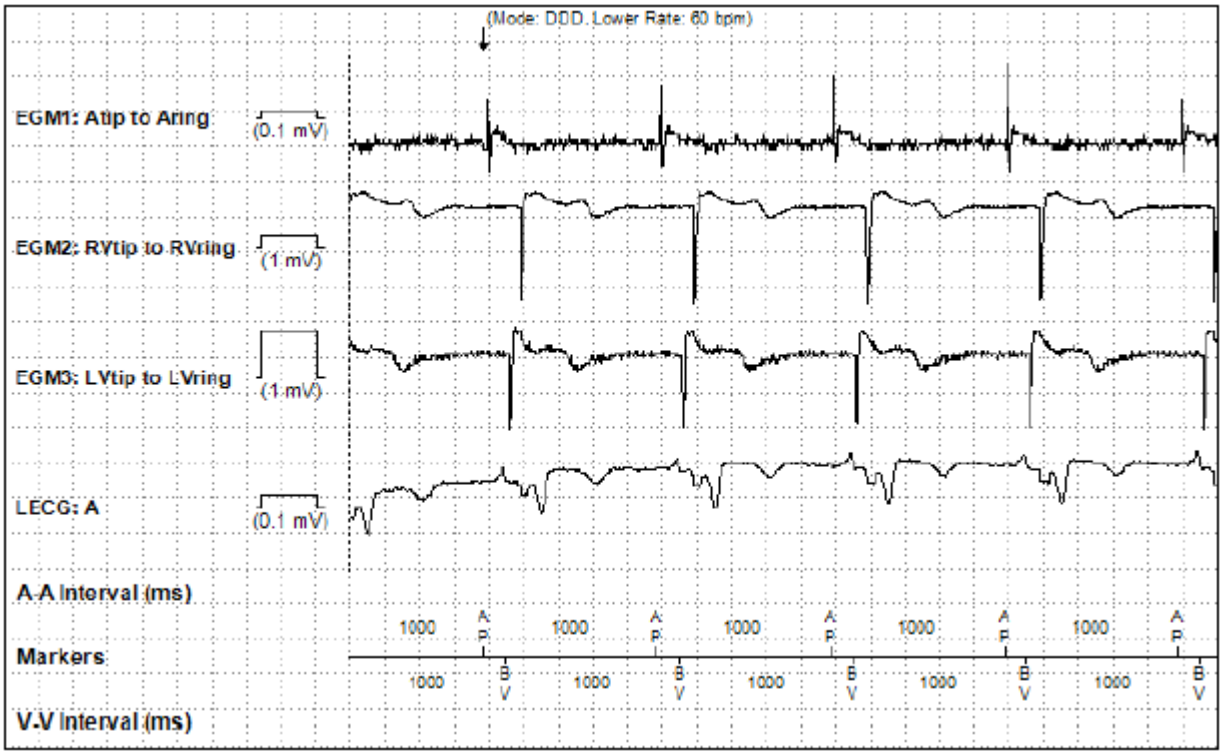

Figure 3: Device EGM showing multisite RV pacing in the RV apex and RV free wall.

At next clinic review the patient reported much improved symptoms and over the next year he had increased exercise tolerance, improvement of his breathlessness and fluid overload allowing reduction in diuretics remaining on just a low dose of Spironolactone and he increased his working hours showing considerable overall improvement to the patient's quality of life.

Discussion

This case study demonstrates an unusual case of ARVC with RVFW disarticulation treated with CRT to resynchronise the right ventricle for relief of right sided heart failure symptoms. Pacing both the RV apex and disarticulated RVFW worked well for this patient giving symptomatic benefit and it may be worth considering for heart failure patients in whom capture in the RVFW can be confirmed.

References

Guiraudon, G. M., Klein, G. J., Gulamhusein, S. S., Painvin, G. A., Del Campo, C., Gonzales, J. C., Ko, P. T. (1983). Total Disconnection of the Right Ventricular Free Wall: Surgical Treatment of Right Ventricular Tachycardia Associated with Right Ventricular Dysplasia. *Circulation*, 67, 2, 463-470.

Hamilton, R. M. (2009). Arrhythmogenic Right Ventricular Cardiomyopathy. *PACE*, 32, Issue S2, S44-S51.

Marcus, F. I., McKenna, W. J., Sherrill, D., Basso, C., Bauce, B., Bluemke, D. A. Calkins, H., Corrado, D., Cox, M. G. P. J., Daubert, J. P., Fontaine, G., Gear, K., Hauer, R., Nava, A., Picard, M. H., Protonotarios, N., Saffitz, J. E., Yoerger Sanborn, D. M., Steinberg, J. S., Tandri, H., Gaetano, T., Towbin, J. A., Tsatsopoulou, A., Wichter, T., Zareba, W. (2010). Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia Proposed Modification of the Task Force Criteria. *Circulation*, 12, 1533-1541.

McKenna, W. J., Thiene, G., Nava, A., Fontaliran, F., Blomstron-Lundgvist, C., Fontaine, G., Camerini, F. et al. (1994). Diagnosis of arrhythmogenic right ventricular dysplasia/cardiomyopathy. Task Force of the Working Group Myocardial and Pericardial Disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology. *British Heart Journal*, 71: 215-218.