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Case Report

Grouped Beating in Eisenmenger: Successful Localization and Ablation of an Accelerated Idioventricular Rhythm from Within the Purkinje System

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Abstract

A 33-year old female with a background of Eisenmenger syndrome secondary to multiple congenital muscular ventricular septal defects (VSD) was admitted with a recent history of frequent intermittent palpitations. It was noted that she had an independent accelerated idioventricular rhythm (AIVR), with rates varying between 85-110bpm, which exhibited a repetitive grouped beating pattern. Although generally perceived as benign, in this case this rhythm was drug refractory, was associated with significant compromise to cardiac filling and output and progressed to haemodynamically intolerable sustained ventricular tachyarrhythmia. Successful ablation was performed at the inferior aspect of the residual VSD, from within the Purkinje network.

Key words: Grouped Beating, Accelerated Idioventricular Rhythm, Eisenmenger syndrome

Case Presentations

A 33-year-old female with a background of Eisenmenger syndrome secondary to multiple congenital muscular ventricular septal defects (VSDs) was admitted with a one-day history of frequent intermittent palpitations. There were no precipitating factors. She was diagnosed with VSDs during childhood and initially underwent pulmonary artery banding followed by surgical VSD repair and pulmonary debanding. However she had residual VSD patency and developed post-operative complete heart block requiring dual chamber pacemaker implantation. She had a previous history of right-sided atrial tachycardia treated with external cardioversion. Her most recent pacemaker interrogation showed atrial flutter and the patient initially underwent a successful cavo-tricuspid isthmus ablation.

At the time of the procedure and post-procedure it was noted that she had an independent accelerated idioventricular rhythm (AIVR), with rates varying between 85-110 bpm (**Figure 1A**) with occasional transient acceleration up to 150 bpm. This AIVR was initially not associated with any symptoms. The cycle length of the AIVR was variable, but exhibited a repetitive grouped beating pattern with occasional progressively shorter RR intervals followed by longer cycles. It had a LBBB morphology suggesting that it had a predominant right sided origin. This rhythm was dissociated from a slower sinus activity and its origin was mapped close to the inferior border of the residual VSD. Attempted ablation of this rhythm was not made initially and medical management was first considered.

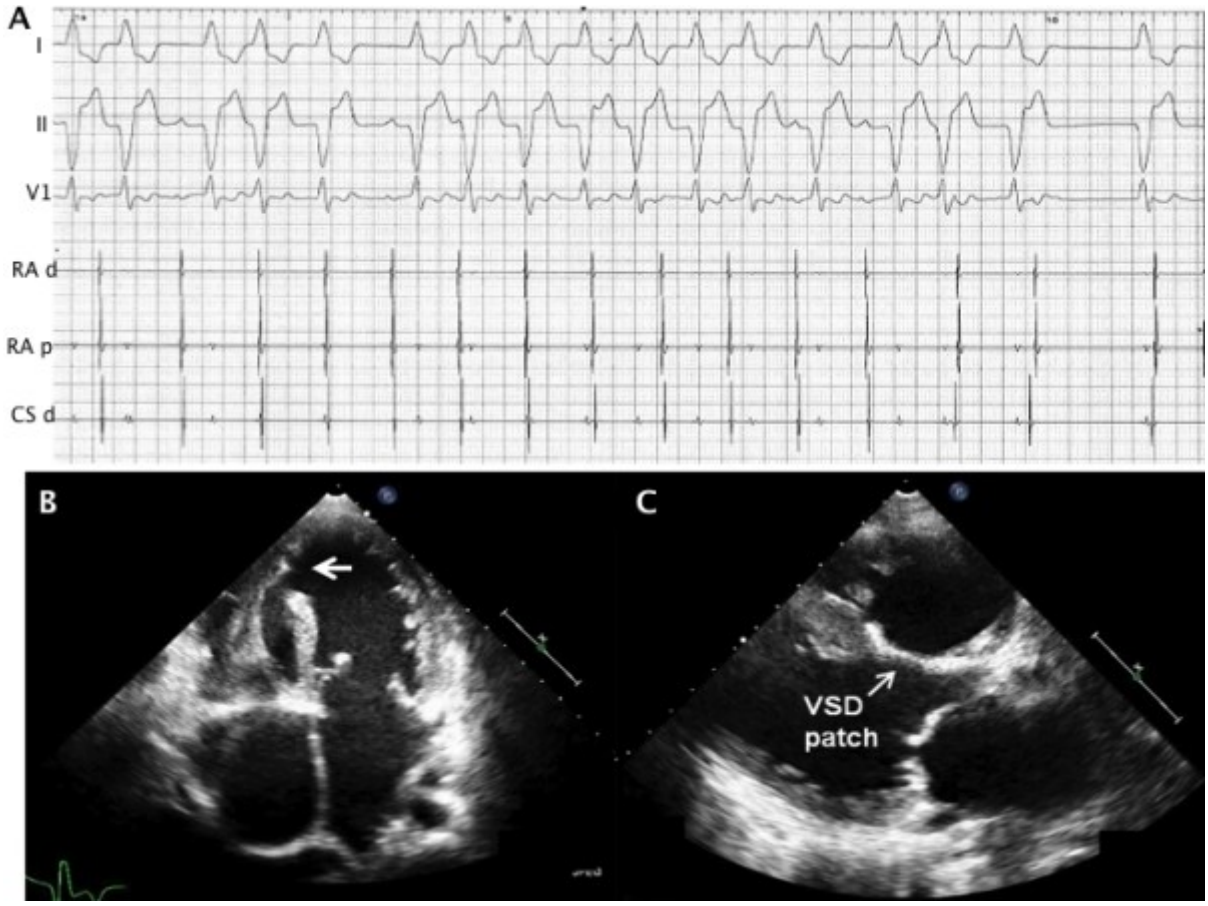

Figure 1: A Tracing recorded at the end of the first ablation procedure for cavotricuspid isthmus-dependent atrial flutter, showing an accelerated idioventricular rhythm with variable cycle length along with corresponding endocavitary signals. Note the presence of ventriculo-atrial dissociation. Paper speed is 25mm/sec. RA d, RA p – right atrial distal and proximal; CS d – coronary sinus distal. **B and C** illustrate transthoracic apical 4 chamber (B) and parasternal long axis (C) echocardiographic views. The apical residual ventricular septal defect is arrowed and measures 1.1cm.

Transthoracic echocardiogram post atrial flutter ablation procedure showed moderately dilated right ventricle with moderately impaired function, severe tricuspid regurgitation, a mean pulmonary artery pressure of 52 mmHg, an apical patent VSD (**Figure 1B**, arrowed) and no residual flow across the previously closed VSD (**Figure 1C**). The left ventricle showed moderate systolic dysfunction with ejection fraction of 43%. Notably, a significantly impaired filling due to reduced atrial emptying during ventricular systole and compromised cardiac output were demonstrated in the presence of frequent ventricular arrhythmia. As there was no significant change to the AIVR with up-titration of beta-blocker and increased atrio-ventricular sequential pacing rate to 90 bpm, along with it being associated with reduced cardiac function, an elective ablation was scheduled. Prior to this however, after having been almost continuously in this rhythm for 5 days without significant associated symptoms, the patient developed sustained ventricular tachycardia (VT) with the same morphology as the