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A hybrid cardioverter-defibrillator implantation approach in a patient with 

Eisenmenger syndrome

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Short title: Hybrid ICD implantation in Eisenmenger syndrome

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Patients with Eisenmenger syndrome (ES) have one of the highest risks of sudden cardiac death (SCD) among patients with congenital heart disease (CHD) [1]. Nevertheless, only a small proportion of ES patients receive implantable cardioverter-defibrillator (ICD) and several issues need to be considered before device implantation.

We present a 71-year-old man with atrial septum defect (ASD) type secundum and permanent atrial fibrillation (AF) who was admitted after cardiac arrest. The first recorded rhythm was ventricular fibrillation (VF) and return of spontaneous circulation was achieved after 8 minutes of resuscitation. Physical examination revealed cyanosis and signs of heart failure. Electrocardiogram confirmed AF and incomplete right bundle branch block. Coronary artery disease and pulmonary embolism were excluded. Echocardiography and computed tomography scan (Figure 1A) showed severely enlarged right atrium and right ventricle (RV) with reduced systolic function, severe tricuspid regurgitation and a large ASD secundum (3.3 cm) with bidirectional shunt. Right heart catheterization confirmed severe pulmonary arterial hypertension. In-hospital monitoring revealed intermittent bradycardia and asystole (maximum 6.5 seconds) without any sustained ventricular tachyarrhythmias.

Secondary prevention ICD implantation was indicated after multidisciplinary team assessment. However, several technical challenges were present due to enlarged right heart chambers, cardiac shunt and bradycardia indication. Transvenous lead placement in patients with cardiac shunts is contraindicated due to high thrombogenicity and risk of systemic embolization [2]. Due to lead survival issues exclusive use of epicardial ICD patches represents a suboptimal solution. Subcutaneous-ICD (S-ICD) is a compelling alternative, however S-ICD lacks bradycardia and anti-tachycardia pacing functions and many CHD patients are not eligible for implantation [3].

Therefore, we decided for a hybrid implantation approach combining a subcutaneous defibrillator lead (6996 SQ 41 cm; Medtronic), an epicardial unipolar defibrillator patch
(6271M oval patch; Medtronic) and an epicardial bipolar pace-sense lead (4968 CapSure Epi 25 cm; Medtronic). The subcutaneous lead was positioned along the right parasternal border using the dedicated tunnelling tool; the epicardial patch was sutured on the posterior left ventricular wall and the epicardial pacing lead was attached in the RV apical region via left mini-thoracotomy. ICD generator (Medtronic Protecta XT DF-1) was implanted in the upper-left abdominal region. Defibrillation vector was programmed between the subcutaneous lead and the epicardial patch (Figure 1B). VF induced via 50 Hz burst was successfully detected and terminated with 20 joules. At 12-months follow-up ICD device parameters were stable and no therapy was delivered.

As shown, complex extravascular device implantation is feasible and safe in carefully selected ES patients. Favourable results have been reported with simultaneous use of S-ICD and epicardial pacemaker in complex CHD patients [4]. Combination of a leadless intracardiac pacemaker and S-ICD could also present a feasible option [5], however it was not considered due to persistent thromboembolic risk and suboptimal integration of both modalities. In complex CHD patients with defibrillation and bradycardia indication we propose a hybrid single-device approach that prevents possible device interactions. Furthermore, epicardial defibrillator patch and subcutaneous lead provide optimization of the defibrillation vector and act as a safety feature in case of dysfunction.
References


Figure 1. Chest computed tomography (CT) scan before and chest radiography after hybrid implantable cardioverter-defibrillator (ICD) implantation in a patient with Eisenmenger syndrome.

Panel A. CT scan demonstrates features of advanced pulmonary hypertension with severely enlarged right atrium (RA), enlarged and hypertrophied right ventricle (RV).

Panel B. Posteroanterior and lateral chest radiographs showing the hybrid ICD device components: (1) subcutaneous defibrillator lead positioned along right parasternal border, (2) epicardial defibrillator patch on the posterior left ventricular wall, (3) epicardial pacing lead in the RV apical region, and (4) ICD generator in the upper-left abdominal region. Arrow indicates the defibrillation vector programmed between the subcutaneous defibrillator lead and the epicardial defibrillator patch.