A hybrid approach to implantable cardioverter-defibrillator implantation in a patient with Eisenmenger syndrome

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

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

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

Considering the above, we decided for a hybrid implantation approach combining a subcutaneous defibrillator lead (6996 SQ 41 cm; Medtronic, Dublin, Ireland), 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 tunneling tool; the epicardial patch was sutured on the posterior left ventricular wall; and the epicardial pacing lead was attached in the right ventricular apical region via left minithoracotomy. An ICD generator (Protecta XT DF-1, Medtronic) was implanted in the upper-left abdominal region. Defibrillation vector was programmed between the subcutaneous lead and epicardial patch (FIGURE 1B). Ventricular fibrillation induced via a 50-Hz burst was successfully detected and terminated with 20 joules. At 12-month follow-up, ICD device parameters were stable and no therapy was delivered.

Our case illustrates that complex extravascular device implantation is feasible and safe in carefully selected patients with Eisenmenger syndrome. Favorable outcomes have been reported for a simultaneous use of S-ICD and epicardial pacemaker in complex cases of CHD.[4] The combination of a leadless intracardiac pacemaker

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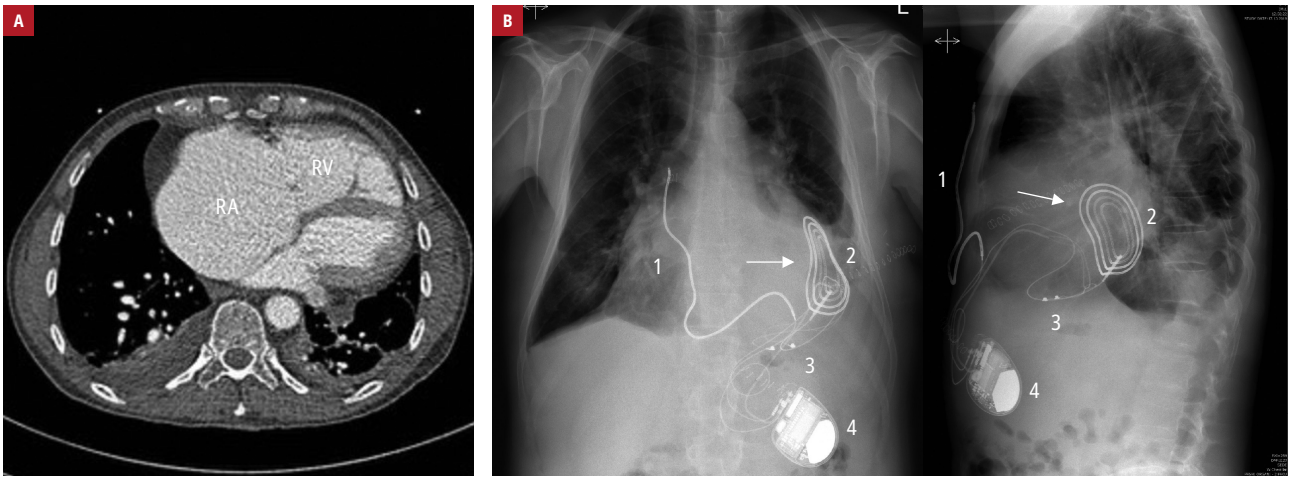

FIGURE 1 Chest computed tomography (CT) before and chest radiography after hybrid implantable cardioverter-defibrillator (ICD) implantation in a patient with Eisenmenger syndrome: **A** – a CT scan demonstrating features of advanced pulmonary hypertension with severely enlarged right atrium (RA) as well as enlarged and hypertrophied right ventricle (RV); **B** – posteroanterior and lateral chest radiographs showing the hybrid ICD device components: 1) subcutaneous defibrillator lead positioned along the 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. Arrows indicate the defibrillation vector programmed between the subcutaneous defibrillator lead and epicardial defibrillator patch.

and S-ICD could also present a feasible option.[5] However, it was not considered in our case due to persistent thromboembolic risk and suboptimal integration of both modalities.

In conclusion, in complex CHD cases with defibrillation and indication for pacing due to bradycardia, we propose a hybrid single-device approach that prevents possible device interactions. Furthermore, an epicardial defibrillator patch and subcutaneous lead provide optimization of the defibrillation vector and act as a safety feature in case of dysfunction.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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