CASE REPORT

Wearable Cardioverter-Defibrillator in a Patient with Left Ventricular Noncompaction/Hypertrabeculation, Coronary Artery Disease, and Polyneuropathy

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A 55-year-old Caucasian male with coronary heart disease was admitted because of dyspnea for 4 weeks. Echocardiography showed a dilated left ventricle with an ejection fraction of 34% and apical left ventricular hypertrabeculation/noncompaction with an apical thrombus. Neurologic examination revealed positional tremor and generally reduced tendon reflexes. During 8 weeks, his condition improved under pharmacotherapy. The patient was skeptical about implantable cardioverter-defibrillator (ICD) and expected further improvement from pharmacotherapy. Thus, he received a wearable cardioverter-defibrillator (WCD). We conclude that a WCD might be useful in noncompaction patients in whom improvement of systolic dysfunction is expected or who are skeptical about ICDs.

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The current ACC/AHA/HRS guidelines mention placement of an implantable cardioverter-defibrillator (ICD) in patients with left ventricular hypertrabeculation/noncompaction (LVHT) as "a reasonable clinical strategy" to reduce the risk of sudden death with a class IIb recommendation, level of evidence C.¹ Application of a wearable cardioverter-defibrillator (WCD) as bridging to ICD-implantation in a LVHT patient has not been reported.²

A 55-year-old Caucasian male was admitted in September 2013 because of increasing dyspnea for 4 weeks. He had a history of arterial hypertension for 5 years, aortic stent-implantation for an infrarenal aortic aneurysm in 2012, and recurrent lumbar pain. He reported that his father had died from stroke and his mother from sudden cardiac death (SCD). Clinical examination showed bilateral pulmonary rales and leg oedema. ECG showed normal sinus rhythm, Q waves in II, III, Avf, and V₁–V₆. Echocardiography showed a

dilated left ventricle with reduced systolic function and an ejection fraction of 34%, applying the biplane Simpson-rule, and LVHT affecting the left ventricular apical segments with a suspected apical thrombus which was confirmed by cardiac magnetic resonance imaging (Fig. 1). Coronary angiography showed occluded left anterior descending and right coronary arteries, filled by collaterals from the circumflex artery which was without stenosis. A neurohumoral therapy with losartan, hydrochlorothiazide, spironolactone and nebivolol, and anticoagulant therapy with phenprocoumon were started. With this therapy, his condition improved and Pro-BNP-levels regressed from 6.218 to 1.489 ng/L. Clinical neurologic examination revealed positional tremor and generally reduced tendon reflexes, why a polyneuropathy was suspected. Nerve conduction studies were normal.

Despite clinical improvement and resolution of the apical thrombus, systolic function in December 2013 still was reduced with a left ventricular

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Figure 1. Echocardiographic apical four-chamber view shows the dilated left ventricle with hypertrabeculation/noncompaction of the left ventricular apex, apical interventricular septum and apical lateral wall. A left ventricular thrombus was suspected in the apex and confirmed by cardiac magnetic resonance imaging.

ejection fraction of 35%, a finding indicating ICD implantation for primary prevention of SCD according to current guidelines. The patient was sceptical about ICD implantation and claimed that probably his cardiac condition and left ventricular ejection fraction would still improve due to the now up-titrated neurohumoral therapy. Thus, we decided to equip him with a WCD and follow-up investigations are scheduled.

LVHT is a cardiac abnormality characterized by a hypertrabeculated left ventricular wall with a sponge-like appearance and a two-layered structure with an outer, subepicardially located compacted zone and an inner, subendocardially located noncompacted zone. LVHT is frequently associated with neuromuscular disorders.³ Cardiac manifestations of LVHT comprise heart failure, thromboembolism, and arrhythmias.⁴ LVHT may be associated with coronary artery disease.⁵ The indication for ICD for primary prophylaxis of SCD in LVHT is a matter of debate. There is increasing evidence that the risk for malignant arrhythmias is associated with decreased systolic function and that guidelines for primary prevention of SCD, developed for patients with dilative or ischemic cardiomyopathy, might also apply to patients with LVHT.⁶

When considering ICD implantation in patients with LVHT, the possible risks and disadvantages of this therapy have to be taken into account:

Complications of ICD in LVHT patients comprise right atrial mural thrombus necessitating warfarin therapy, hemothorax after epicardial placement of the lead necessitating prolonged mechanical ventilation, inadvertent lead positioning within the cardiac vein and inappropriate discharges due to atrial arrhythmias.⁷ Furthermore, magnetic resonance imaging studies will be impossible in ICD patients.

When LVHT with left ventricular dysfunction is diagnosed in a previously untreated patient, it is uncertain if neurohumoral therapy will improve his systolic function and thus ICD implantation will be necessary in the future at all. A WCD (LifeVest, ZOLL, Pittsburgh, PA, USA) is a useful tool for bridging the time until the patient has stabilized and neurohumoral therapy has been up-titrated.² Our patient presented with the combination of ischemic cardiomyopathy, systolic dysfunction, and LVHT. Although the systolic function was severely depressed at initial presentation, his condition improved under neurohumoral therapy and he expected a further improvement why he denied immediate ICD implantation. A WCD was chosen to bridge the time until either the left ventricular function might have improved or—in the worst case—the patient has agreed with ICD implantation in the case no improvement of systolic function could be achieved.²

We conclude that WCD may be a useful option in patients with LVHT and systolic dysfunction in whom improvement of systolic dysfunction under neurohumoral therapy might be expected or in whom the patient is still skeptical about ICD implantation.

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