Cardiac devices and neuromuscular disorders in left ventricular noncompaction

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Left ventricular hypertrabeculation/noncompaction (LVHT) is associated with arrhythmias (Table 1) and neuromuscular disorders (NMD) [1–4]. Cardiac implantable electronic devices (CIED) like antibradycardic pacemakers (PM), implantable cardioverters/defibrillators (ICD) and biventricular pacemakers for cardiac resynchronization (CRT) are beneficial but have also risks [5–7]. The aim of the study was to analyze the indications, complications and outcome of LVHT-patients with CIED. Included were all patients in whom LVHT was diagnosed since 1995 [2]. There was no prespecified protocol, patients were treated by several physicians, and CIED were implanted by different institutions. In May 2010, the records were screened. Date and indications for implantation, survival status, type of CIED, complications and current pharmacotherapy were recorded [8,9].

Included were 154 patients (28% female, mean age 53 ± 16, range 14–94 years) with LVHT. The baseline data are listed in Table 2. In May 2010, 40 (27%) of the 154 patients were no longer alive. Causes of death were heart failure (n = 11), pneumonia (n = 6), hepatic failure (n = 4), sudden death (n = 4), malignancy (n = 3), sepsis (n = 3), stroke (n = 3), pulmonary embolism (n = 2), renal failure (n = 1) or unknown (n = 3). Of the 4 patients who died suddenly, one had received a CRT-system 7 years before, one did not consent to the proposed ICD, and two deaths occurred in the 1990s in patients who had not received CIED but presumably would receive ICD for primary prevention today. During 65 months (1–167), the annual mortality was 4.81%.

CIED were implanted in 24 patients (16%). Patients with CIED had more often heart failure, left-bundle-branch-block, valvular abnormalities and LVHT affecting the lateral wall, larger left ventricular enddiastolic diameters and a poorer left ventricular systolic function than patients without CIED (Table 2).

ICD were implanted in 15 patients (Table 3). Coronary angiography was normal in 13/15 patients. ICD were implanted for primary prevention in 9 and for secondary prevention in 6 patients. ICD were combined with CRT in 8 patients. In two further patients a CRT-system was implanted 7 and 2 years after the ICD. In patient 16, CRT-implantation was unsuccessfully attempted 7 years after the ICD. During 47 months (6–170) after implantation, one or more appropriate discharges occurred in three patients. In two of them (4 and 14) ICD were implanted because of primary, and in patient 5 as secondary prevention. Device-related complications occurred in 5 patients: lead fracture (5 and 16), hematothorax (3), pericardial effusion with lead malposition (6) and septicemia (13) [11]. During follow-up, one

Table 1
Arrhythmias reported in patients with left ventricular hypertrabeculation/noncompaction [3].

Arrhythmia	Children	Adults
Atrial fibrillation, n	1	95
Atrial flutter, n	1	3
Supraventricular tachycardia, n	6	18
Ventricular pre-excitation via accessory pathways, n	24	18
Wolff–Parkinson–White syndrome, n	24	17
Mahaim fibers, n	0	1
Atrioventricular block, n	24	31
First-degree atrioventricular block, n	4	14
Second-degree atrioventricular block, n	4	0
Complete atrioventricular block, n	16	17
Sick sinus syndrome, n	5	5
Atrial standstill, n	1	1
Bradycardia, n	15	3
QT prolongation, n	5	42
Ventricular tachycardia, n	17	118
Ventricular fibrillation, n	9	5

Table 2
Baseline characteristics of 154 patients with left ventricular hypertrabeculation/noncompaction, with and without cardiac implantable electronic devices.

Characteristic	All patients (n = 154)	No CIED (n = 130)	CIED (n = 24)
Age, years	53	53	54
Female, %	28	29	25
Observation, years	5	5	6
Neurologically normal, %	10	12	4
Specific neuromuscular disorder, %	14	13	21
Neuromuscular disorder of unknown etiology, %	46	44	54
Neurologically not investigated, %	30	32	21
Diabetes mellitus, %	18	19	17
Arterial hypertension, %	39	42	25
No heart failure, %	33	38	4†
Heart failure			
NYHA I, %	8	9	4
NYHA II, %	14	12	25
NYHA III, %	27	24	42*
NYHA IV, %	19	18	25
No ECG abnormality, %	11	13	0
Left-bundle-branch-block, %	20	12	58†
Atrial fibrillation, %	17	15	29
Left ventricular enddiastolic diameter, mm	62	60	72†
Left ventricular fractional shortening, %	23	25	16†
Valvular abnormalities, %	58	53	88*
Location of LVHT			
Apex, %	94	95	92
Anterior wall, %	3	4	0
Posterior wall, %	16	16	13
Lateral wall, %	49	44	75*
Interventricular septum, %	1	2	0
Sum of affected cardiac regions, n	1.6	1.6	1.8
Deceased during follow-up, %	26	25	33

*p < 0.010.
†p < 0.001.
CIED = cardiac implantable electronic devices.
LVHT = left ventricular hypertrabeculation/noncompaction.

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Table 3
LVHT-patients with implantable cardioverters and/or cardiac resynchronization therapy.

Number/year of birth/sex	Indication	Type/Impl	LVHT	LVFS	AF	NMD	FU	Pharma	Arrh, CIED-compl
1/1948/m	HF/PP	CRT/ICD/2008	2010	10%	Yes	NI	3	BB, Diu, Digi, ACEI; heart transplantation 02/10	Short ventricular runs
2/1935/f	SP	ICD/2001	2006	17%	Yes	U	2	Deceased 05/06 of heart failure	None
3/1958/m [10]	HF/SP	CRT/ICD/2004	1998	14%	No	U	98	Deceased 01/06 of pneumonia	Hematothorax at implantation
4/1944/m	HF/PP	CRT/ICD/2006	2007	20%	No	U	24	ATIIB, BB, Diu, OAC	Multiple shocks due to ventricular tachycardia
5/1958/m [10]	HF/SP	ICD/1994 CRT/2001	1998	11%	No	D	107	ACEI, BB, OAC	Multiple shocks due to ventricular tachycardia, inappropriate shocks due to ICD-lead fracture
6/1940/m [11]	PP	ICD/2007	2007	28%	No	U	36	ATIIB, Diu	Pericardial effusion, malpositioned ICD lead in cardiac vein, inappropriate shocks
7/1962/m [10]	HF/PP	CRT/ICD/2002	1995	11%	No	D	87	Deceased 10/02 of septicemia	None
8/1931/m [10]	HF	CRT/2001	2006	11%	Yes	U	20	Deceased 01/08 of sudden death	None
9/1978/m[a]	HF/SP	DDDR/2004 ICD/2005 CRT/2007	2006	18%	Yes	U	12	Deceased 12/07 of heart failure	None
10/1967/f	SP	ICD/2007	2008	24%	No	NI	15	BB, ATIIB	None
11/1935/f [10]	HF/PP	CRT/ICD/2006	1999	25%	No	N	131	ACEI, BB, Diu, ASA, Dig	Short ventricular and atrial runs
12/1923/f [10]	HF	CRT/2001	1996	17%	No	D	163	ACEI, BB, Diu, ASA	Surgery because of pericardial effusion
13/1954/m	HF/SP	CRT/ICD/2006	1999	13%	No	NI	122	Deceased 06/09 of septicemia	Septicemia 2 months after implantation, inappropriate shocks due to AF
14/1951/m [10]	HF/PP	CRT/ICD/2003	1998	7%	No	U	86	OAC, ACEI, digitalis, Diu, BB, Amio	Multiple shocks due to ventricular tachycardia and ventricular fibrillation
15/1969/m	HF/PP	CRT/ICD/2009	2009	14%	No	NI	3	ACEI, BB, Diu, ASA	None
16/1945/m	HF/PP	ICD/2001	2001	15%	Yes	U	118	Amio, ACEI, BB, Diu, Dig, OAC	Nonsustained ventricular tachycardia; fracture of right ventricular lead; placement of left ventricular lead impossible due to no adequate cardiac vein
17/1965/m	PP	ICD/2008	2006	8%	No	U	41	ACEI, ivabradine, BB	None
18/1933/f [10]	HF	CRT/2001	2000	6%	No	U	119	ACEI, BB, Diu, Dig	Short ventricular and atrial runs

ACEI = angiotensin converting enzyme inhibitors.
AF = atrial fibrillation.
Amio = amiodarone.
Arrh = arrhythmias.
ASA = acetylsalicylic acid.
BB = betablockers.
CIED-compl = device-related complications.
CRT = cardiac resynchronization device.
D = definite neuromuscular disorder.
Dig = digitalis.
Diu = diuretics.
FU = months since LVHT diagnosis.
HF = heart failure.
ICD = implantable cardioverter defibrillator.
Impl = year of CIED implantation.
LVFS = left ventricular fractional.
LVHT = year of LVHT diagnosis.
N = normal neurologic examination.
NI = not investigated.
NMD = neuromuscular disorder.
OAC = oral anticoagulation.
Pharma = pharmacotherapy in May 2010.
PP = primary prevention of cardiac death.
SP = secondary prevention of cardiac death.
U = neuromuscular disorder of unknown etiology.
[a] This patient is also listed in Table 4.

patient underwent heart transplantation and 5 patients died due to heart failure (n = 2), septicemia (n = 2) and pneumonia (n = 1).

Three patients received CRT-systems, and in 10 patients it was combined with ICD (Table 3). Five of the patients who received a CRT-system died due to septicemia (7 and 13), heart failure (9), pneumonia (3) or suddenly at home (8). The condition of patient 1 deteriorated despite CRT and he underwent cardiac transplantation. The remaining 7 patients improved and were classified as CRT-responders. Patients with atrial fibrillation (AF) who received a CRT either died (8 and 9) or underwent cardiac transplantation (1). A further patient (13) developed AF and aggravation of heart failure and died due to septicemia.

In 7 patients a PM was implanted because of symptomatic bradycardia (Table 4). Coronary angiography in 5 of them showed coronary artery disease in two patients (23 and 24). Heart failure was present in all patients and was the cause of death in two patients (9 and 23). Three of the seven patients had died until May 2010. In two of the 4 surviving patients (19 and 22) it was uncertain if symptoms were caused by heart failure or NMD. Two patients (20 and 21) responded well to pharmacotherapy. In none of the patients, heart failure was attributed to right ventricular pacing. Insufficient frequency-control aggravated heart failure in patients 9, 20 and 22.