

Cardiac pacing in 21 patients with Emery-Dreifuss muscular dystrophy: a single-centre study with a 39-year follow-up

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Abstract

Background: Emery-Dreifuss muscular dystrophy (EDMD) is a genetic condition associated with cardiac arrhythmias. The patients typically develop early, asymptomatic bradyarrhythmia, which may lead to sudden death, preventable with a cardiac implantable electronic device (CIED). EDMD may be characterised by atrial electrical silence. Intra-operative electrophysiological evaluation of the myocardium helps ultimately determine the true nature of the disorder and select an appropriate CIED.

Aim: To analyse permanent electrotherapy procedures in EDMD patients: atrial pacing limitations that stem from the electrophysiological properties of the myocardium and long-term follow-up of implanted devices.

Methods: A total of 21 EDMD patients (mean age 29 ± 9 years) with a CIED implanted (1976–2014) due to bradyarrhythmia were included in the study. The implantation procedures and factors determining the CIED type selection were analysed.

Results: CIEDs were implanted in five women and in 16 men with EDMD types 1 and 2 (mean follow-up: 11 ± 8 years). Intra-operatively assessed atrial electrophysiology resulted in changing the planned CIED type during the procedure in three men with EDMD type 1. Eventually, we implanted: eight DDD, one VDD, 11 VVI, and one CD-DR device, with four of the patients' devices switched later from DDD to VVI mode in response to electrophysiological changes in the atria.

Conclusions: Intra-operative assessment of atrial electrophysiological properties resulted in changing the planned DDD mode for VVI in 19% of patients with EDMD type 1. Progression of the underlying disease over a 39-year follow-up resulted in a later change of the initially selected pacing mode from DDD to VVI in 40% of cases.

Key words: Emery-Dreifuss muscular dystrophy, cardiac arrhythmias, conduction abnormalities, cardiac pacing

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INTRODUCTION

Emery-Dreifuss muscular dystrophy (EDMD) presents with neurologic symptoms and cardiac problems that may be life-threatening as well as difficult to diagnose. Neurologically, the disease manifests as humeroperoneal muscle weakness and atrophy, joint contractures, and spinal stiffness. These factors determine the phenotype. EDMD is associated with structural and/or functional deregulation of genes encoding proteins that create the nuclear envelope [1, 2]. Although EDMD patients have similar phenotypes, the genes respon-

sible for the disease may be different. EDMD type 1 patients have a mutation in emerin (*EMD*) located on the X chromosome (Xq28) [3]. EDMD 1 is an X-linked recessive disease. Women do not have skeletal muscle symptoms; however, 20% may have cardiac symptoms. EDMD type 2 patients have mutations in the *LMNA* gene, which encodes lamin A and C. It is located on chromosome 1 (1q22) [4]. However, in almost 60% of EDMD patients, mutations are found in other genes that encode nuclear proteins.

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The clinical presentation of EDMD is exceptional because even mild pathology of the skeletal muscles is accompanied by cardiac pathology, which may result in sudden cardiac death (SCD) [5]. The first cardiologic symptoms are observed during the second to fourth decades of life. Symptoms can be mild or even absent, which seems to be the reason behind SCD in EDMD patients [6]. In the beginning of disease development, the atria are affected, while the size of the left ventricle remains unaffected for a long period of time. The first symptoms include conduction block, bradyarrhythmia, and/or tachyarrhythmia. Early diagnosis and implantation of a permanent pacemaker protects against SCD, but it does not mitigate disease progression [7, 8]. The heart can be affected prior to the muscular system. Ventricular arrhythmias are the main cause of SCD [9]. An implantable cardioverter-defibrillator (ICD) should be implanted for primary prevention of these arrhythmias [10–12].

Fatty and fibrous infiltrates can be found in the myocardium and the cardiac conduction system. Morphologically, this results in electrical inhomogeneity. Inhomogeneous degeneration affects various cardiac areas, including the atria and ventricles [5, 13]. The EDMD-associated changes that develop over time in the nervous and cardiovascular systems have both direct and indirect effects on preliminary indications for, and the ultimate implantation of, the specific permanent electrotherapy device.

We present the results of cardiac implantable electronic device (CIED) placement in EDMD patients throughout a 39-year follow-up. The procedures were performed by one operator (RS). Most papers describe only a few cases. Rarely, larger groups of patients are presented [14, 15].

The aim of our study was to analyse permanent electrotherapy in EDMD patients. We focused on: (i) atrial pacing limitations due to electrophysiological properties of the myocardium and (ii) long-term follow-up of implanted devices.

METHODS

Patients

Twenty-one patients with EDMD type 1 or 2 (mean age at the time of procedure was 29 ± 9 years) were hospitalised between December 29, 1976 and December 31, 2013 for elective CIED implantation. The follow-up periods lasted from six months to 36 years (mean: 11 ± 8 years).

All patients showed a complete EDMD phenotype (Table 1). EDMD type 1 (*EMD* gene) GenBank accession no. NM000117. EDMD type 2 (*LMNA* gene) GenBank accession no. NM170707.

Twelve-lead electrocardiogram (ECG) at rest and/or 24-h Holter monitoring in these patients revealed significant dysfunction of the cardiac conduction system, which was an indication for CIED.

Device implantation procedure and follow-up

Transvenous implantations of CIEDs were performed as usual: in the subclavicular region, with local anaesthesia. Leads were

inserted through the cephalic and/or axillary or subclavian vein. As EDMD patients need to turn to one side in order to assume an upright position and in the process tend to use the arm less affected by the disease, the side of device implantation was selected individually, based on where it would least inhibit the patient's mobility.

Leads were placed under fluoroscopy. Specific electrical measurements were analysed with the appropriate device. Lead placement depended on the parameters obtained during a pacing test, lead tip stabilisation (passive or active), and electrophysiological properties of adjacent tissue recorded during the procedure.

Atrial leads were placed in the right atrial appendage. Ventricular leads were placed in the right ventricle at the site with optimal pacing parameters: the lowest pacing threshold and a voltage amplitude that ensured proper functioning of the device; this was intended to avoid areas of myocardial inhomogeneity.

Patients with preserved spontaneous sinus rhythm underwent atrioventricular node conduction assessment after atrial lead placement but prior to any ventricular lead insertion. In patients with suspected atrial electrical silence, atrial stimulation and pacing parameters were assessed in various areas of the heart.

After discharge, patients were followed-up in the outpatient clinic. We interrogated the devices and, when needed, modified the pacing mode.

RESULTS

We included patients undergoing their first implantation procedure (six females, mean age: 38 ± 11 years; 15 males, mean age: 25 ± 7 years; $p = 0.016$). The mean follow-up period was 10.6 ± 7.9 years. Sixteen patients had EDMD type 1 (mean age: 26 ± 8 years), and five patients had EDMD type 2 (mean age: 36 ± 11 years); EDMD type 1:EDMD type 2, NS ($p = 0.068$).

All of the patients (those with sinus rhythm as well as those with atrial fibrillation [AF]) had various degrees of atrioventricular conduction block. In patients with ECG tracings at rest showing normal sinus rhythm and normal atrioventricular conduction, intraprocedural atrial stimulation at an increased rate revealed nodal dysfunction (Table 1). Nearly 30% of ECG tracings showed intraventricular conduction anomalies: left anterior fascicular block or incomplete right bundle branch block.

Mild or even asymptomatic bradyarrhythmia was common, and it was independent of the rhythm shown in ECG tracings (sinus rhythm, junctional rhythm, or chronic AF). We observed bradyarrhythmia at a rate of 20 bpm in 24-h Holter monitoring. Nonetheless, they did not result in symptomatic central nervous system ischaemia. No patient had fully symptomatic Morgagni-Adams-Stokes syndrome prior to device implantation. Patient symptoms included irregular heartbeat and palpitations. There were episodes of paroxysmal AF (Table 1).

Table 1. Emery-Dreifuss muscular dystrophy (EMMD) patient characteristics in chronological order of procedures. Genetic mutations and cardiac arrhythmias recorded prior to the procedure

Pt. No.	Sex	Follow-up (months since procedure)	Dystrophy type	Form: FF/SF	Gene mutation	Age during procedure	Rhythm before/during procedure	AVB/WP during procedure	PACs	PVCs according to L-W	CIED (planned) implemented/ → retained
1	M	1976/432 † 01.2013	EDMD 1	FF	c.256C>T, p.Gln86X	22	CAF	+		II	VVI
2	M	1996/210	EDMD 1	FF	c.187+1G>A	34	SR/SAB	III	PAF	0/1	VVI
3	M	1996/209	EDMD 1	FF	c.153delC, p.Ser52AlafsX13	21	Atrial standstill	+ (junctional rhythm)		0/1	(DDD)VVI
4	M	1996/193 † 12.2012	EDMD 1	FF	c.153delC, p.Ser52AlafsX13	28	SR/SAB	I	Isolated	0/1	VVI
5	M	1997/198	EDMD 1	FF	c.153delC, p.Ser52AlafsX13	34	CAF	+		0/1	VVI
6	M	2000/102 † 01.2009	EDMD 1	SF	c.397C>T, p.Gln133X	25	CAF	+		II	VVI
7	M	2000/162	EDMD 1	FF	c.153delC, p.Ser52AlafsX13	25	SR < 40/min		SVT	0/1	DDD → VVI
8	M	2001/152	EDMD 1	FF	c.153delC, p.Ser52AlafsX13	32	SR/SAB	I/II	Isolated	II	DDD → VVI
9	M	2001/146	EDMD 1	SF	c.153delC, p.Ser52AlafsX13	18	CAF	+		II	VVI
10	M	2001/145	EDMD 1	FF	c.1A>G	24	SR < 34/min	WP < 130	Isolated	0/1	DDD
11	F	2002/50 † 2006	EDMD 2	FF	c.1357C>T, p.Arg453Trp	41	CAF	+		0/1	VVI
12	M	2002/135	EDMD 1	FF	c.450dup, p.Glu151GlyfsX59	18	SR/SAB	I/II/III	Isolated	0/1	DDD → VVI
13	F	2002/133	EDMD 2	SF	c.1072G>A, p.Glu358Lys	25	SR	I/II	Isolated	0/1	VDD → VVI
14	F	2003/10 † 2004	EDMD 2	FF	c.788T>C, p.Leu263Pro	28	CAFL	+		0/1	VVI
15	M	2005/97	EDMD 1	FF	c.399+1G>C	16	SR/SAB	WP < 130		0	DDD
16	M	2007/78	EDMD 1	SF	c.417_418dup, p.Leu140PhefsX98	18	SR	SAB WP < 130	PAF	0/1	DDD

↑

Table 1. cont. Emery-Dreifuss muscular dystrophy (EMMD) patient characteristics in chronological order of procedures. Genetic mutations and cardiac arrhythmias recorded prior to the procedure

Pt. No.	Sex	Follow-up (months since procedure)	Dystrophy type	Form: FF/SF	Gene mutation	Age during procedure	Rhythm before/during procedure	AVB/WP during procedure	PACs	PVCs according to L-W	CIED (planned) implemented/ → retained
17	M	2008/67	EDMD 1	FF	c.187+1G>A	35	Atrial standstill	+ (junctional rhythm)		0/1	(DDD)VVI
18	F	2008/66	EDMD 1	FF	c.187+1G>A	44	SR	II	Isolated	0/1	DDD
19	M	2009/57	EDMD 1	SF	c.1 A>G	25	Atrial standstill	+ (junctional rhythm)		0/1	(DDD)VVI
20	F	2011/28	EDMD 2	FF	c.1355T>C, p.Val452Ala	52	SR	I/II	Isolated	II	DDD
21	M	2013/6	EDMD 2	SF	c.1357C>T, p.Arg453Trp	35	SR	I		IVb	ICD-DR

EDMD type 1 (gene *EMD*) GenBank accession no. NM000117; EDM2 type 2 (gen *LMNA*) GenBank accession no. NM170707; †Date of death; AVB — atrioventricular block; CAF — chronic atrial fibrillation; CAFL — chronic atrial flutter; CIED — cardiac implantable electronic device; F — female; FF — familial form; L-W — Lown's grade of premature ventricular contractions; M — male; PACs — premature atrial contractions; PAF — paroxysmal atrial fibrillation; PVCs — premature ventricular contraction/complexes; SAB — sinoatrial block; SF — sinoatrial rhythm; SR — sinus rhythm; SVT — supraventricular tachycardia; WP — Wenckebach point (< 130)

Table 1 presents the devices we had planned to implant, the devices we actually implanted, and the associated cardiac arrhythmias registered prior to, or during, the implantation procedure. We planned to implant a VVI device in six patients with atrioventricular conduction abnormalities, AF, atrial flutter, or atrial tachycardia insufficiently controlled with medication. Fifteen patients had atrioventricular conduction abnormalities in ECG and 24-h Holter monitoring. Two patients had atrial electrical silence. After periprocedural Wenckebach point analysis, four patients with sinus rhythm had atrioventricular conduction ratio of 1:1, at the rate not faster than 130 impulses per minute (mean 120 imp/min). In these four patients we implanted a device for atrioventricular pacing. One patient developed AF during the procedure. After a failed attempt to manage the arrhythmia with drugs, we implanted a VVI pacemaker. Because of atrial electrophysiological properties noted during the procedure in three patients, we implanted a VVI pacemaker instead of a DDD device that we had planned to implant (Table 1). Because of abnormalities at the atrioventricular node, we did not implant any AAI pacemakers. The device pocket was located in the left subclavian region in nine patients (in the right subclavian region in 12 patients). The pocket was situated over the pectoral muscle.

Presentation of cases

The indication for pacemaker implantation in a 21-year-old patient (PA, no. 3), was a complete atrioventricular block with ventricular rhythm of 36 bpm. During the procedure, performed on July 15, 1996, 100-impulse/min pacing with a lead placed in the right atrial appendage resulted in a significant delay (190 ms) between the pacing spike and atrial excitation (Fig. 1).

A 35-year-old patient (DG, no. 17) received a pacemaker due to bradycardia (38/min from the atrioventricular node). The procedure was performed on May 15, 2008. Due to lack of excitability of the atrial myocardium, we changed the primary device type that had been planned for implantation. Lack of excitability was consistent in all of the tested places in the right atrial endocardium and veins leading to the coronary sinus; it also did not depend on the scale of electrical impulse.

The indication for the procedure in a 25-year-old patient (DR, no. 19), was atrioventricular node rhythm (42/min). A VVI pacemaker was implanted on March 30, 2009. Due to lack of excitability of the right atrium in response to electrical impulses, no lead was placed in this heart chamber. The scale of electrical impulse had no effect on cardiac excitation.

During the procedure in a 41-year-old patient (KG, no. 11), we observed actual electrophysiological properties of the atrium (Fig. 2).

We found low-amplitude atrial flutter and excluded atrial electrical silence suggested by earlier ECG and Holter findings (Fig. 3).

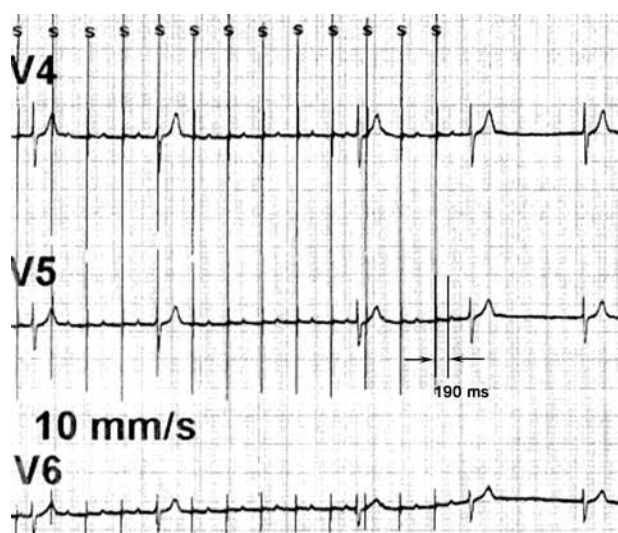


Figure 1. Delay (190 ms for pacing rhythm 100 imp/min) between pacing spike and atrial excitation

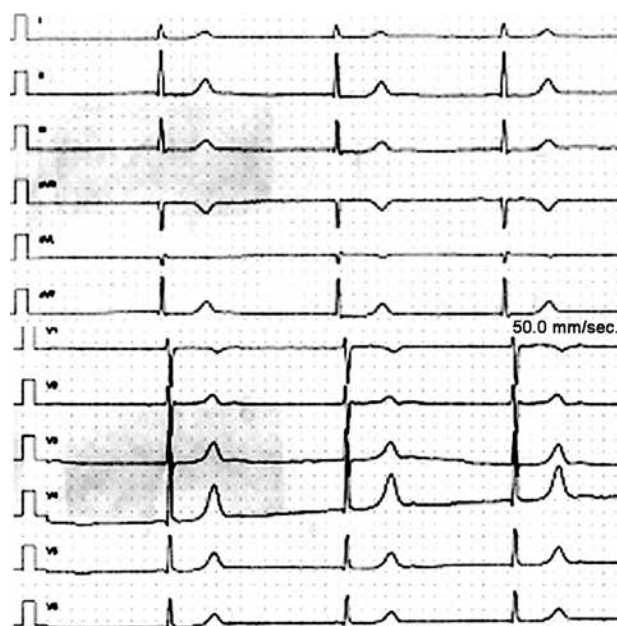


Figure 2. Patient no. 11, 12-lead electrocardiogram at rest. Initially, we interpreted it as a nodal rhythm of 42/min and sinus arrest or complete atrioventricular block. No atrial electrical activity

During lead implantation (screw-in, bipolar) in patient DG (no. 17), prior to final lead placement, we found cardiac areas of a markedly increased pacing threshold. An magnetic resonance imaging scan showed an area of delayed enhancement in the right ventricle, suggesting fibrosis.

During follow-up, three patients had an additional operation due to pacemaker dysfunction (no planned pacemaker replacement).

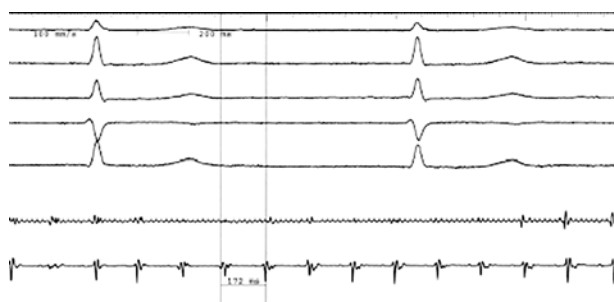


Figure 3. The same patient during an electrophysiological study. Figure is from the lead in the right atrium; atrial flutter

One month after the implantation procedure in a 21-year-old patient, PA (no. 3), unsuccessful pacing was observed in the ventricular lead (tined, unipolar) placed in the right ventricular apex (pacing threshold > 4.8 V, 0.5 ms; initially: 0.9 V, 0.5 ms). Because of a suspected exit block, glucocorticoids were prescribed, which proved ineffective. A new lead (tined, bipolar) was implanted in a new place, where the pacing threshold was also increased (1.9 V, 0.5 ms).

Patient DA (no. 1) received a new VVI mode pacemaker 20 years after the first procedure. The indication for the second procedure was lead failure and no intrinsic rhythm.

An examination conducted three months after the procedure in patient DA (no. 2) revealed no pacing due to Twiddler's syndrome. A chest radiogram showed spiral torsion of the lead in the pocket and right ventricle. The unipolar lead in the adapter area was completely broken. We replaced the adapter and inserted the pacemaker into a new pocket, achieving adequate pacing.

Pacemaker interrogations in the rest of the patients showed parameter changes (including pacing threshold values) to be consistent with those normally occurring during and after post-implantation healing. The generated pulse amplitude (voltage) in patients with a preserved intrinsic rhythm with a rate above the minimal rate programmed into the device did not change in the follow-up period in an outpatient setting.

Six of the 10 patients received a DDD pacemaker. In four out of these patients the pacemaker mode was changed to VVI 3–7 years (mean 5 ± 1.6 years) after the first procedure. The indication for that change was chronic AF. One male patient with EDMD type 2 received an ICD-DR because of unsustained ventricular tachycardia, clinical presentation, and genetic background. This decision was made according to the SCD primary prevention guidelines.

Five patients died (three had EDMD type 1, two had EDMD type 2). Autopsies were not performed. Three patients (no. 1, 4, and 6) had SCD; one had ischaemic stroke (no. 11); and one had advanced heart failure (no. 14). These patients had been paced for a mean of 13.2 ± 24.8 years (range: 1–36 years).

DISCUSSION

EDMD is a congenital myopathy. The life-threatening aspect of the condition, however, is not due to the slowly progressive neurological anomalies, but to cardiac arrhythmias, which depend on disease stage [7, 15–17].

These bradyarrhythmias are mildly symptomatic or even asymptomatic. This is not related to the aetiology of bradyarrhythmia, instead it seems to be a result of patient adaptation to slower and slower rhythm during significantly decreased physical activity compared with that of their peers. The asymptomatic character of bradyarrhythmia was an important obstacle in obtaining patient consent for the procedure. Supraventricular arrhythmias were present in patients with EDMD type 1 and 2 [18]. Most of the patients had chronic AF or atrial flutter. Moreover, in some of the patients treated with atrioventricular pacing and antiarrhythmic medications, AF progressed to its chronic form. In some patients with EDMD, cardiac pacing indicated based on ECG tracings may not be possible to implement. In some of these patients, electrical instability, which facilitates AF, might be present with decreased cardiac tissue excitability. The mechanism and grade of atrial failure influences the electrophysiological properties observed during the procedure and affects the steps of the procedure [10, 15].

Electrical atrial silence is typical for EDMD. Patients with that electrophysiological condition require a VVI pacemaker implantation or reprogramming of a pre-existing CIED into a VVI mode. Electrical atrial silence heralds a decline in atrial haemodynamic function [18, 19]. Electrical silence shown in a surface ECG trace needs to be verified during the procedure, as was done in this case.

At our institution, during three implantations in patients with EDMD type 1, unfavourable atrial electrophysiology precluded the initially planned atrial pacing. One of those patients developed a considerable atrial spike-to-P-wave delay. This may have been associated with hyperpolarisation of excitable tissue because of degeneration, which led to increased conduction time. It seems that the other two patients had similar electrophysiological mechanisms, but more advanced and with complete failure to conduct pacemaker-generated impulses.

Most of our patients were in their second to fourth decade of life, which is consistent with other publications [4, 9]. We observed conduction abnormalities, as well as an increased chance of supraventricular arrhythmias in people in that age range [20]. We triggered supraventricular arrhythmias during atrial lead placement in DDD pacemaker implantation procedures. Then, we observed short episodes of AF or atrial flutter, which resolved with no treatment. However, AF triggered in one patient during the procedure could not be interrupted with pharmacological treatment. Therefore, we implanted a VVI pacemaker.

Because of the progression from paroxysmal to chronic AF, we changed the pacing mode to VVI in four patients during follow-up. To our knowledge, the data presented here constitute the greatest number of EDMD patients who received a DDD pacemaker and were followed up for one of the longest periods. There is no published data in this field that can be compared with our results. For example, a study by Boriani et al. [18] included 18 patients; 10 out of these patients received pacemakers: nine VVI/VVIR and one DDDR. The median follow-up after pacemaker implantation was 12 years. Pacemaker dependency was recorded in all cases at the end of follow-up. Five (50%) implanted patients developed atrial standstill after the development of paroxysmal AF or atrial flutter and one patient developed permanent AF after pacemaker implantation.

The atria and ventricles are involved in heterogenic degeneration of the myocardium in which muscular tissue is replaced by fibrotic and adipose tissue [5, 10, 13]. In all patients except one leads were placed in areas with proper pacing parameters despite the possibility of local right ventricular degeneration.

The type and progression of arrhythmia were similar in all of our patients. However, what we could actually observe in individual patients during follow-up were the various stages of arrhythmia progression. Thanks to these observations, we managed to identify the chronology of electrophysiological states of the atria: 1) isolated supraventricular beats, 2) sustained arrhythmia evident in surface ECG, 3) sustained arrhythmia, detectable only during periprocedural intra-atrial assessment of electrophysiological properties in patients with surface ECG tracings suggesting atrial electrical silence, and, eventually, 4) true atrial electrical silence.

In summary, our study shows limitations of pacemaker implantation caused by atrial electrical silence and long-term changes observed during follow-up. Despite generally similar outcomes, we observed differences in terms of arrhythmia onset and detectability via surface ECG. In light of this observation, our research appears to be one of the few studies presenting subsequent stages of electrophysiological changes in the heart, particularly in the atria. These changes include early development of atrial rhythm abnormalities, which can be diagnosed using ECG. Some of the atrial rhythm abnormalities were diagnosed during periprocedural electrophysiological studies when the surface ECG tracing suggested electrical silence. In some cases, there was true electrical silence. We also presented genetic mutations in patients who underwent electrotherapy.

Limitations of the study

Over a period of almost 40-year experience with patients with EDMD at our centre, the indications for CIED have changed. In some patients with an implanted pacemaker,

the pacing mode should be modified according to current American College of Cardiology/European Heart Rhythm Association guidelines. However, progression of cardiovascular abnormalities is not as inconvenient for EDMD patients as the neurological aspect of their condition. It seems to be the reason for rejection of device adjustment by EDMD patients.

Device materials and design have changed over the study period. In the beginning, passive leads determined the pacing site in the right ventricular apex. Screw-in leads overcame this problem, and now it is feasible to pace from other sites.

In patients with DDD-mode pacing systems, we also assessed other right atrium sites besides the auricle, and the obtained results were similar. However, even having screw-in leads at our disposal, we still ended up placing them in the auricle because our earlier observations indicated good long-term pacing results from that location in other EDMD patients.

A retrospective analysis of electrophysiological measurements performed today vs. results of earlier studies does not allow for drawing unequivocal conclusions in this area, due to differences in the devices and methods used to collect data.

CONCLUSIONS

Because of the electrophysiological atrial properties measured during the procedure, we had to implant VVI instead of the previously planned DDD pacemakers in 19% of patients with EDMD type 1. Because of the progression of the underlying disease in the atria and rhythm disturbances during a 38-year follow-up, we had to change the double-chamber (atrial and ventricular) pacing to ventricular pacing in 40% of cases. Atrio-ventricular blocks of various degrees were seen in all patients.

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Conflict of interest: none declared

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Stymulacja serca u 21 chorych z zespołem dystrofii mięśniowej Emery'ego-Dreifussa: jednośrodkowe badanie z 39-letnią obserwacją

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Streszczenie

Wstęp: Dystrofia mięśniowa Emery'ego-Dreifussa (EDMD) jest genetycznie uwarunkowaną jednostką chorobową, której rozwojowi towarzyszą zaburzenia rytmu serca. Fazę początkową cechują bradyarymie, zazwyczaj bez objawów klinicznych, prowadzące do nagłych zgonów w sytuacji braku zabezpieczenia stałą stymulacją serca. Specyficzną cechą jest występowanie „ciszy elektrycznej” przedsionków, przy czym dopiero śródzabiegowa ocena właściwości elektrofizjologicznych miokardium pozwala na weryfikację jej rzeczywistego charakteru i implantację odpowiedniego układu.

Cel: Celem pracy była analiza zabiegów stałej elektroterapii serca u pacjentów z EDMD w aspekcie ograniczeń i możliwości wdrożenia określonego trybu stymulacji serca i jego utrzymania podczas dalszej obserwacji.

Metody: Badanie dotyczyło 21 chorych z EDMD (śr. wieku $28,6 \pm 9,4$ roku), którym w latach 1976–2014 z powodu bradyarymii powodowanych dysfunkcją układu bodźcowo-przewodzącego serca implantowano urządzenia do stałej elektroterapii serca. Analizie poddano przebieg zabiegów i czynniki determinujące implantację określonego typu stymulatora.

Wyniki: Urządzenia implantowano u 5 kobiet i 16 mężczyzn z EDMD typu 1 i typu 2 (śr. czas obserwacji: $10,6 \pm 7,9$ roku). Właściwości elektrofizjologiczne tkanki przedsionków wykryte śródzabiegowo wpłynęły na zmianę rozważanego do wdrożenia typu stymulacji u 3 mężczyzn z EDMD typu 1. Ostatecznie implantowano układy: 8 — typu DDD, 1 — typu VDD, 11 — typu VVI, 1 — typu ICD-DR. Zmiany elektrofizjologiczne zachodzące w przedsionkach wpłynęły na zmianę programu DDD do trybu VVI u 4 osób.

Wnioski: Elektrofizjologiczne właściwości tkanki przedsionków określone śródzabiegowo ograniczyły możliwość wdrożenia planowanej stymulacji z DDD do VVI u 19% chorych z zespołem EDMD typu 1. Progresja procesu podczas 39-letniej obserwacji wpłynęła na konieczność zmiany stymulacji typu DDD na VVI w 40% przypadków.

Słowa kluczowe: dystrofia mięśniowa Emery'ego-Dreifussa, stymulacja serca, zaburzenia rytmu i przewodzenia

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