Non-compaction cardiomyopathy – complications and long-term outcomes: a single-center experience

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Aim: To investigate prevalence of arrythmias and thromboembolic events, as well as long term outcomes among patients diagnosed and treated of non-compaction cardiomyopathy (NCC) according to current recommendations in University Hospital Centre (UHC) Zagreb.

Patients and Methods: A single center retrospective study was conducted. Patients newly diagnosed with NCC in UHC Zagreb during period 2009-2018 were analyzed. The diagnosis was confirmed by both echocardiography and cardiovascular magnetic resonance. Hospital database and charts were used for clinical data, echocardiography data was obtained from digital database using EchoPac. Patients were followed-up clinically and by the means of echocardiography.

Results: 32 patients (pts), 18 men (53.25 %) were included. At the time of diagnosis (baseline), mean age was 47.7±15.4 years, majority of pts (84.38% of pts, N=27) were in functional NYHA class ≥2, with mean NT-proBNP values of 3870±6619 ng/L. Echocardiography revealed reduced left ventricular systolic function; baseline ejection fraction (EF) was 27.52±11.94%. Patients were discharged with heart failure therapy: beta-blockers (30 pts, 93.75%), angiotensin-converting enzyme inhibitors or angiotensin receptor blockers (29 pts, 90.63%), angiotensin receptor–neprilysin inhibitor (1 pts, 3.13%), mineralocorticoid receptor antagonists (28 pts, 84.38%); and 26 pts (81.25%) required symptomatic diuretic use. Cardiac resynchronization therapy with defibrillator was implanted in 11 pts (34.38%) and implantable cardioverter defibrillator (ICD) in 16 pts (50%). At baseline, 9 pts (28.13%) were already receiving anticoagulation due to previous thromboembolic events (**Table 1**). At discharge, 17 pts (53.13%) were anticoagulated (warfarin in 14 pts, 82.35%, novel direct oral anticoagulants in 3 pts, 17.65%). Mean follow-up period was 3.42±3.31 years. At the end of follow up period, functional improvement was observed: 21.88% (N=7) pts were in NYHA ≥2 status (p<0.05), with manifest, but statistically nonsignificant reduction of NT-proBNP levels (1260±2266 ng/L, p=0.063). Control echocardiography revealed significant improvement in EF (40.24±11.39%, p<0.001). There were no new thromboembolic events. While arrythmias were common at the time of diagnosis (**Table 1**), there was only one ICD activation during follow-up. None of the pts have died, 1 patient received heart transplant and 2 were implanted with left ventricular assist device.

Conclusion: Optimal medical treatment in patients with NCC[1] corresponds with good long-term outcomes, functional improvement, and low risk of recurrent thromboembolic events or malignant arrythmias.

TABLE 1. Number of patients diagnosed with thromboembolic events and arrythmias at the time of diagnosis and at the end of the follow-up period.

Number of patients:	At diagnosis	At follow up
THROMBOEMBOLIC EVENTS		
Left ventricular thrombus	3 (9.38%)	0 (0)
Stroke/TIA	4 (12.50%)	0 (0)
Other embolus	1 (3.13%)	0 (0)
ARRYTHMIAS		
Atrial fibrillation	3 (9.38%)	7 (21.88%)
nsVT	15 (46.88%)	8 (25.00%)
Sustained VT	1 (3.13%)	4 (12.50%)
ICD activation	0 (0%)	1 (6.25%)

TIA = transient ischemic attack; nsVT = non-sustained ventricular tachycardia; VT = ventricular tachycardia; ICD = implantable cardioverter defibrillator.

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LITERATURE

1. Carrilho-Ferreira P, Almeida AG, Pinto FJ. Non-compaction cardiomyopathy: prevalence, prognosis, pathoetiology, genetics, and risk of cardioembolism. Curr Heart Fail Rep. 2014 Dec;11(4):393-403. https://doi.org/10.1007/s11897-014-0227-3