Abstract: **P1810**

The superimposed myocarditis in arrhythmogenic right ventricular cardiomyopathy: the role in the course of the disease and the results of treatment

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Purpose: to study the role of myocarditis in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC), and to evaluate the results of its treatment.
Methods: 54 patients (38.7±14.1 y., 42.6% men) with ARVC according to Revised Task Force Criteria 2010 were evaluated (34 patients with definite, 18 - with borderline, 2 with possible diagnosis). Follow-up period 21 [6; 60] months. All patients underwent ECG, 24h-Holter monitoring, echocardiography, DNA-diagnostic, blood tests for detection of anti-heart antibodies (AHA) and DNA of viruses. Also were performed cardiac MRI (n=49), signal-averaged ECG (n=18), endomyocardial biopsy (n= 2), autopsy (n=2).
Results: myocarditis was diagnosed in 38 (70.4%) patients, 8 of whom were virus-positive (2 by myocardium, 6 by blood). Immunosuppressive therapy (IST) was conducted in 25 patients and included hydroxychloroquine (n=22, 200 mg/day), steroids (n=7, 16 [8; 24] mg/day), azathioprine (n=2, 150 mg/day). Patients with myocarditis who received and not received IST were compared. Initially, patients receiving IST had a longer term of disease, higher titers of AHA and a larger end diastolic volume of the left ventricle (LV) by MRI. Patients who received IST, had significantly lower mortality in comparison with patients with myocarditis without treatment (4.0 vs 30.8%, p=0.03). Only patients with myocarditis treated with IST demonstrated significant positive dynamics in PV?s number (11,7 [2,6; 37] vs 0,8 [0,01; 4.5] thousand/day., p<0.001); nonsustained ventricular tachycardia (VT, 57.9 vs 26.3%, p=0.034); sustained VT (SVT, 31.6 vs 0%, p=0.014); their LV ejection fraction (EF) remained stable (52.1±14.8 vs 52.4±13.5%, p=0.58). In patients with myocarditis without IST, there was a tendency to EF reduction (64,3±8,8 vs 57,2±9,4, p=0,058). Comparison of patients with isolated ARVC and ARVC plus myocarditis revealed no differences in structural and functional parameters (severity of arrhythmias, EF of both ventricles, heart chambers size, functional class of CHF, etc.), in the effectiveness of radiofrequency ablation and in the frequency of adverse outcomes. The absence of differences is regarded as the result of effective IST. However, myocarditis was significantly less common in patients with the most typical form of ARVD (SVT without significant CHF; with mutations in the PKP2 gene) than in patients with latent arrhythmic form (without SVT, with mutations in the DSG, SCN5A, FLNC genes) and with biventricular ?HF (mutations in DSP, DES genes).
Conclusion: the frequency of superimposed myocarditis in patients with ARVC exceeds 70%. Myocarditis in ARVC could be primary (including viral) or secondary (autoimmune). Regardless to etiology, myocarditis in ARVC should be actively diagnosed and treated, because patients with myocarditis not receiving IST have significantly worse effectiveness of antiarrhythmic therapy and outcomes in comparison with patients with ARVD and myocarditis, received IST.