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Echocardiographic study in children suffering from Familial Mediterranean fever . A preliminary study of a cohort of patients from Northern Greece.

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Objectives: Familial Mediterranean Fever (FMF) is anautoinflammatory periodic disease, inherited in an autosomal recessive manner. It is characterized by recurrent febrile episodes and sterile serosal inflammation attacks.Echocardiographic abnormalities (systolic or diastolic dysfunction) have been described in a few studies, which were related with sustained subclinical inflammation. The aim of the present study was to assess cardiac function in children diagnosed with FMF as well as the correlation of echocardiographic parameters with the disease duration,inflammatory markers [Erythrocyte Sedimentation Rate (ESR), CRP] and the serum amyloid levels.

Methods: 24 patients with FMF (12 males,12 females,mean age: 8.64±3.55 years) during attack-free period and 18 healthy controls (7 males,11 females, mean age:9.51 ± 2.78 years) were enrolled in this study, excluding children with obesity, hypertension and dyslipidemia. All FMF patients fulfilled the diagnostic Tel Hashomer criteria and were under colchicine treatment. The disease was confirmed by genetic analysis. A single experienced paediatric cardiologist recorded all echocardiographic measurements using conventional echocardiography.

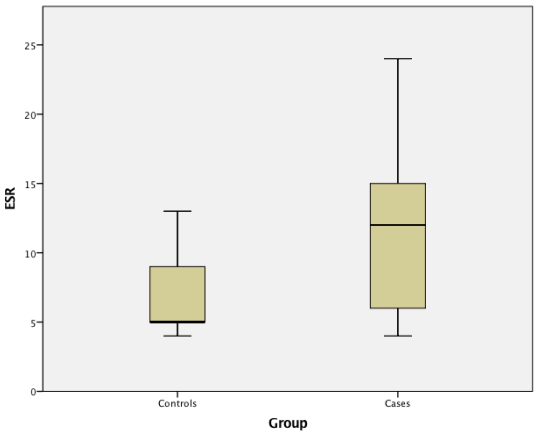

Figure 1. ESR by group. Significantly higher for Cases.

Results: Mean disease duration of the study population was 5.88±2.97 years. Serum amyloid levels in FMF patients ranged between 0.7 and 128 mg/L(normal values < 6.4mg/L). Although ESR and CRP values where within normal limits, mean values of ESR were statistically significant increased in patients compared to controls (11.39±5.74 vs 6.72±2.56, p=0.003).The echocardiographic study did not yield statistically significant differences in the parameters of systolic and diastolic function (table). Furthermore, no statistical significant correlations were found between echocardiographic parameters, inflammation markers and amyloid levels.

	FMF	Controls	p
Age (years)	8.63 ±3.54	9.5 ±2.78	0.392
ESR(mm)	11.39 ±5.74	6.72 ±2.56	0.003
CRP(mg/dl)	0.49 ±0.65	0.21 ±0.19	0.088
LVIDd (mm)	39.1 ± 6.4	41.8 ±6.6	0.189
LVIDs (mm)	24.6 ± 0.41	26.1 ±4.1	0.232
LVPWd (mm)	6.6 ± 1	6.7 ±1.1	0.762
IVSd (mm)	6.5 ±1	6.5 ± 1	0.918
EF%	67.4 ±5.05	67.7 ±5.67	0.835
FS%	37 ±3.97	37.44 ±4.46	0.736
LAD (mm)	25.6 ±3	26.2 ± 2.6	0.529
LA/Ao	1.26 ±0.21	1.24 ±0.1	0.681
E wave MV (m/sec)	0.85 ±0.11	0.81 ±0.07	0.165
A wave MV(m/sec)	0.45 ± 0.11	0.45 ±0.07	0.913
dt MV (ms)	131 ±35.16	133.77 ±21.78	0.770
E/A MV	1.96 ±0.43	1.84 ±0.27	0.305
E wave TR (m/sec)	0.72 ±0.1	0.74 ±0.1	0.421
A wave TR (m/sec)	0.45 ±0.05	0.45 ±0.1	0.974

Conclusions: Children suffering from FMF that are under colchicine treatment do not display early echochardiographic abnormalities. This could be related to the young age, the early diagnosis along with the short duration of the disease. Moreover, colchicine probably protects patients from the harmful effects of autoinflammation on myocardial tissue. However, FMF children should be followed with echocardiography as they are considered to have an increased risk of cardiac function abnormalities.