

(female gender, hypertension, diabetes, and smoking) and did not find any statistically significant effects ($p>0.05$ each) on adverse outcomes except for increased rates of postoperative atrial fibrillation (POAF) in patients with diabetes ($p=0.03$).

The authors have also stated that they find the lower rates of POAF in obese group very conflicting. But, as mentioned in the original article, obese patients are more prone to insulin resistance which mandates increased use of perioperative insulin for strict blood glucose control. Insulin causes decrease in the occurrence rates of POAF (2). We find this explanation for the lower rates of POAF satisfactory.

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Noncompaction with dysmorphism, mental retardation, general wasting, and hypogonadism requires neurologic and sophisticated cytogenetic investigations

To the Editor,

We read with interest the article published entitled "Case of fatal heart failure with biventricular noncompaction, genital skeletal abnormalities and mental retardation." by Ataş et al. (1) regarding a 48-year-old female from consanguineous parents with dilated cardiomyopathy (dCMP), left ventricular hypertrabeculation/noncompaction (LVHT), primary amenorrhea, bilateral amazia, ovarian dysgenesis, uterine aplasia, hypergonadotropic hypogonadism, macrocephaly, facial acromegaly, arachnodactyly, pectus carinatum, and mental retardation who died from heart failure 4 months after being diagnosed with LVHT. We have the following comments and concerns.

We do not agree with the statement that LVHT is a genetic disorder. Although LVHT is associated with various monogenic disorders, in particular neuromuscular disorders (NMDs) and cardiomyopathies, and chromosomal defects (2), a causal relation between these genetic defects and LVHT has not yet been proven. The strongest argument against a causal relation is that only a small number of patients with

NMDs, cardiomyopathies, and chromosomal defects present with LVHT (2). An argument in favor of a causal relation, however, is that LVHT also occurs familial (3).

The patient underwent cytogenetic investigation; however, it is not mentioned which technique was applied (1). Did the authors investigate complex chromosomal re-arrangements and micro-aberrations by means of fluorescence in-situ-hybridization (FISH) or microarray assays? In particular, did they apply multi-color FISH, telomere/subtelomere FISH, reverse painting, fiber FISH, quantitative FISH, or cobra-FISH?

According to Figure 1, the patient presented with generalized muscle wasting (1). Was this due to being bedridden prior to admission or was this due to involvement of the peripheral nerves or the skeletal muscles? Did the patient ever undergo a clinical neurologic investigation, nerve conduction studies, or needle electromyography? This is of particular importance because LVHT is associated with NMDs in more than half of the cases.

Concerning mental retardation and macrocephaly, it would be interesting to know cerebral imaging results. Was there cerebral atrophy, calcification, demyelination, or hydrocephalus? Did she ever develop seizures? Was an electroencephalogram ever recorded?

Because LVHT can be complicated by stroke embolism, it is important to understand whether the individual or family history was positive for stroke/embolism. Did cerebral imaging reveal previous embolic stroke? Furthermore, patients with LVHT and dCMP require oral anticoagulation with vitamin-K antagonists for primary prophylaxis of stroke/embolism (4). Did the patient receive phenprocoumon or warfarin in addition to heart failure therapy on dismissal?

Furthermore, because LVHT is complicated by arrhythmias, it would be worthwhile to know the results of long-term electrocardiography recordings. Did the two sisters and brother who deceased in childhood die suddenly? Was the family history positive for falls, syncope, fainting, or sudden cardiac death? Was an autopsy conducted in the three deceased children?

Because LVHT may be acquired in some cases (5), it would be interesting to know whether the patient had undergone previous echocardiographies and if these were revised for LVHT?

Overall, this interesting case merits further evaluation with regard to genetic background and possible neuromuscular or cerebral comorbidities. Only if LVHT patients are comprehensively investigated, the pathogenetic background of this enigmatic cardiac abnormality may be elucidated.

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Author's Reply

To the Editor,

We would like to thank you for your criticism in this issue to our paper published in the *Anatol J Cardiol* (1). We appreciate the comments and want to briefly address the main questions raised in your letter. Noncompaction cardiomyopathy (NC) represents heterogeneity in its genetic pattern, pathophysiologic findings, and clinical presentations (2). The American Heart Association classified this entity as a primary genetic cardiomyopathy (3). According to the World Health Organization and European Society of Cardiology classification of cardiomyopathies, NC is still an unclassified cardiomyopathy (3-5). Additionally, there are several reports stating NC as genetic disorder and explain its inheritance and genetic cause (5). Because the laboratory investigations revealed hypergonadotropic hypogonadism and a pelvic MRI demonstrated the absence of ovaries, uterus, or prostate in our patient, we performed conventional cytogenetic analysis to identify whether any chromosomal abnormalities may be associated with these extra cardiac manifestations. Cytogenetic analysis demonstrated a 46, XX karyotype without any chromosomal abnormalities. We did not perform other techniques to investigate complex chromosomal re-arrangements and micro-aberrations. Techniques, such as FISH, CGH, and microarray, may identify the likely genetic etiologies. After evaluation of all the cardiac and extracardiac manifestations, dysmorphic signs, and pedigree analysis, we investigated the most probable candidate gene LMNA mutations associated with cardiomyopathies. Direct sequencing did not reveal any mutations in the coding region of the LMNA gene. To identify the genetic cause of NC in our patient, other known genes associated with NC should be investigated.

The patient had generalized muscle wasting since the first hospitalization. It was most probably associated with heart failure. We referred the patient to neurology during the first admission, and cerebral MR was performed; however, it did not reveal cerebral atrophy, calcification, demyelination, or hydrocephalus. There was suspicion for microangiopathic vascular involvement. Nerve conduction studies or needle electromyography was not performed. Due to non-adherence to the medical treatment, there were recurrent hospitalizations with heart failure decompensation; however, ischemic stroke, seizures, or syncope was not observed. According to cerebral MR findings, there was no sign suggesting previous stroke(s). Because of mental retardation and non-adherence to medical therapy, oral anticoagulant therapy was not administered.

To exclude any arrhythmia, we monitored the patient with telemetry during hospitalization and performed 24-h rhythm Holter but did not detect any arrhythmia. The patient had three healthy brothers and a sister, two sisters and a brother had suddenly died in childhood from unknown rea-

sons; however, an autopsy was not conducted. A brother and sister of the patient were examined by echocardiography; however, there were no abnormality. Rest of the family members were considered as normal.

In conclusion we did not detect any finding, suggesting neuromuscular disease in our evaluation. Coexistence of biventricular NC, genital and skeletal anomalies, and mental retardation led us to consider the presence of a syndrome.

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Thrombus formation during septal puncture

To the Editor,

We deeply appreciate Bilge et al. (1) for this study published in September 2014 issue of *The Anatolian Journal of Cardiology* entitled "Left atrial spontaneous echo contrast and thrombus formation at septal puncture during percutaneous mitral valve repair with the MitraClip system of severe mitral regurgitation: a report of two cases." It was reported in both cases that activated clotting time (ACT) of patients were higher than 250 s; however, it was not emphasized whether unfractionated heparin (UFH) was administered before or after septostomy. This issue is important in patients, particularly with atrial fibrillation (AF) due to risk of thrombus formation. We have reported a case of mitral stenosis and AF who was administered UFH after septostomy and developed thrombus right after trauma of puncture of interatrial