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Letter to the Editor

Noncompaction Necessitates Comprehensive Cardiac and Neurologic Investigations

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With interest we read the article by Espinola-Zavaleta *et al.*, (2006) on the cardiac findings and outcomes in 53 patients with left ventricular hypertrabeculation / noncompaction (LVHT) (Espinola-Zavaleta, N. *et al.*, 2006). Their findings and conclusion raise the following concerns.

The frequency of 74% isolated LVHT is higher than previously reported (Stöllberger, C., & Finsterer, J. 2004). How to explain this unexpected figure? Was it due to the age distribution of the 53 patients or is LVHT more easily overlooked in patients with congenital cardiac disorders?

According to previous reports the right ventricle is hypertrabeculated in the majority of the cases. How to explain that almost two thirds of the cases had a normal number of trabeculations in the right ventricle? How was normality of right ventricular hypertrabeculation defined?

The authors report a relation between the extension of LVHT and the left ventricular function. Was there also a positive and significant correlation between the magnitude of LVHT and fractional shortening or ejection fraction?

It is important to mention that LVHT is not exclusively a congenital abnormality. In single cases with neuromuscular disorders LVHT developed during life-time (Finsterer, J. *et al.*, 2006). For that reason previous echocardiographic examinations should be reviewed in all patients with LVHT. In how many of the presented patients were previous echocardiographs reviewed?

Were measurements of LVHT parameters carried out at end-systole or end-diastole? According to Oechslin's definition they should be carried out at end-systole (Oechslin, E. N. *et al.*, 2000).

It is mentioned that LVHT was mixed up with dilative cardiomyopathy, restrictive cardiomyopathy, congenital heart disease, or heart valve disease. How did the authors exclude that LVHT did not coexist with any of these disorders?

Which was the cause of pericardial effusion in the 3 patients mentioned?

How to explain that all segments carring LVHT were hypokinetic? Is it due to increased wall thickness or decreased elasticity of the left ventricular myocardium?

How many of the patients underwent coronary angiography? Which was the result of coronary angiography in the 11 patients who complained about chest pain?

Which were the risk factors for atherosclerosis or embolism in the three patients who experienced a thromboembolic event?

Chin *et al.* were not the first who described LVHT. LVHT was initially described by Engberding (1984).

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LVHT is not only associated with Barth syndrome but with a number of other neuromuscular disorders and other rare genetic disorders, including dystrobrevinopathies, zaspopathies, myoadenylate-deaminase deficiency, myotonic dystrophy, dsytrophinopathy, hereditary neuropathy, Pompe's disease, or Friedreich's ataxia. Thus, LVHT is not only associated with mutation in the FKB12, CSX, G4.5, or DTNA (dystrobrevin) gene but also in the dsytrophin, cypher/ZASP, lamin A/C, GAA, DMPK, AMPD1, mitochondrial, frataxin, or PMP22 genes (Finsterer, J. *et al.*, 2006; Kanemoto, N. *et al.*, 2006). Because of the frequent association of LVHT with neuromuscular disorders, all patients in whom LVHT is detected should also undergo comprehensive neurological investigations.

Since the pathogenesis of LVHT is still unknown more extensive studies on the etiology of this rare cardiac abnormality are warranted. This includes the development of animal models also to study the long-term risk and prognosis of these patients and to develop supportive measures.

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