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Letter to the Editor Open Access

QT-prolongation and noncompaction

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Letter to the Editor

In a recent article, Szulik et al. reported about a 22 years old female with ventricular fibrillation, QT-prolongation, and left ventricular hypertrabeculation/noncompaction (LVHT) who died from hypoxic cerebral damage 5 days after admission [1]. We have the following comments and concerns. Patients with LVHT have a disposition for any type of cardiac arrhythmia [2,3]. This is why ventricular fibrillation not only could be due to hereditary long-QT syndrome but also due to LVHT. Ventricular fibrillation was either due to LVHT or a consequence of QT-prolongation. QT-prolongation is not unusual in LVHT and has been reported in several cases (table 1) [3-7]. LVHT has been also reported in association with long-QT-syndrome due to mutations in the KCNQ1 gene [8], in the KCNH2 gene [9], or due to an unidentified genetic defect (table 1) [10]. In a study of 105 patients with LVHT, the QT-interval increased during a mean follow up of 3.6y in 15 patients and normalized in 21 patients [11]. The increase was associated with the extension of LVHT and the presence of a neuromuscular disorder (NMD) [11].

Table 1: Patients with LVHT and QT-prolongation

Patient	AA	Mutated gene	Treatment	Reference
Aquired QT-prolongation				
Infant	WPW	None	Drugs	[13]
Female	VF	None	ICD	[4]
Newborn	AVB	None	PM, CRT	[5]
Female, 12yo	VT, HF	None	Drugs	[6]
Female, 22y	SCD	None	Drugs	[1]
Female, 40y	AVB, VT	None	PM, drugs	[7]
47 patients	Nm	None	Nm	[3]
Hereditary long-QT syndrome				
Female, 5y	CA	KCNQ1	Drugs	[8]
Newborn, male 5y	VT, LVHT	KCNH2	Drugs	[9]
Newborn	VT	Nm	PM, ICD	[10]

AA: additional abnormality, WPW: Wolff-Parkinson-White syndrome, VF: ventricular fibrillation, AVB: AV-block, VT: ventricular tachycardia, HF: heart failure, CA: cardiac arrest, LVHT: noncompaction, PM: pace maker, CRT: cardiac resynchronization therapy, ICD: implantable cardioverter defibrillator, Nm: not mentioned

Since LVHT is associated with NMD in up to 80% of the cases [12], it would be interesting to know if the patient presented with any clinical features of a NMD or if the individual or family history was positive for a NMD. Additionally, it is of interest if the mother or any other relative had developed features of an NMD or had been diagnosed previously with a NMD. Second most frequently, LVHT is associated with chromosomal abnormalities. Was there any indication for numeric or structural chromosomal abnormalities? Since LVHT as well as sudden cardiac death (SCD) occurs familiarly, it would be interesting to know if the family was screened for cardiac disease after SCD of the index patient's mother. In case the index patient had undergone echocardiography at the time of her mother's decease, was LVHT detected already at this investigation? Were other family members screened for LVHT or ventricular arrhythmias? Did the index patient ever report palpitations, dyspnoe, leg edema, or exercise intolerance before the mother's decease? Did the index patient undergo an echocardiographic investigation prior to her mother's decease? Since LVHT is not only associated with arrhythmias and heart failure but also with stroke or embolism, it is also of interest if the index patient ever experienced an embolic stroke or peripheral embolism prior to the lethal event. Did her deceased mother ever experience a cardioembolic problem prior to SCD? Overall, this interesting case confirms that LVHT may be also associated with QT-prolongation and that associated ventricular arrhythmias may be fatal in some of these patients. In patients with SCD, relatives should be screened for LVHT and for severe arrhythmias. Implantation of an ICD should be considered in patients with LVHT and a family history positive for SCD. Treatment of OT-prolongation in LVHT depends on the cause, cardiac comorbidity, and duration of QT-prolongation.

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