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Reply

We are very thankful to Russo et al. for their useful comments.[1] We hope that these comments and our answers may further clarify which factors may affect different frequency of the left ventricular dysfunction observed in different series of patients with myotonic dystrophy type 1 (DM1).

Russo and colleagues[1] reported that frequency of the left ventricular dysfunction is higher in studies by Bhakta et al., Petri et al., Groh et al. and Tanawutiiwat et al. than in our study although mean age and gender distribution were similar in all cohorts.[2–6] However, it is of note that other authors did not report neither on the age at onset (the disease form) nor on duration of the disease at the moment of examination, thus we cannot say that these studies comprise entirely similar DM1 populations. Further on, patients in our study were selected consecutively during their first hospitalization, while in other studies known DM1 patients were invited for the examination[2–4] and it is possible that those who were doing bad were more likely to come on follow-up creating a selection bias.

Another important issue is that in our study only standard echocardiography was used, while Bhakta et al. and Groh et al. used different neuroimaging methods (echocardiography, nuclear imaging, contrast ventriculography) including only the most abnormal measurements in the final analysis.[2,4]

It is true that percentage of atrial arrhythmias in our cohort is small, but again only standard 12-lead ECG was used, while in some other studies results from the 24 h Holter ECG monitoring[3] or a history of atrial arrhythmias[2,4] were included, increasing sensitivity and finding higher percentage of arrhythmias, as expected. Further on, we must note that frequency of atrial arrhythmias reported in the Letter to the Editor by Russo et al.[1] is actually given at follow-up for some studies cited. Reported frequencies are lower at the study entry. For instance, in study by Bhakta and coworkers atrial arrhythmias were present in 19 of 205 males and seven of 201 females making overall percentage of 6.4%, not 22.7%. In study by Groh et coworkers,[4] atrial tachyarrhythmia based on the historical data was present in 21 of 406 subjects (5.1%) and based on ECG only six of 406 (1.4%), not 12.8%. Accordingly, our results of 1.8% are based on ECG at the first visit to the referral neuromuscular center. This is also the reason why no one had pacemaker or ICD implanted.

Left ventricular bundle branch block was present in nine (8.1%) DM1 patients in our study. Presence of LBBB was not in association with diastolic dysfunction of the left ventricle. On the other hand, systolic dysfunction was present in 22.2% of patients with LBBB and only 3.9% of those without LBBB ($p < 0.05$). This is in line with previous researches published by Russo and colleagues that showed association between LBBB and left ventricular dysfunction/heart failure.[7,8]

In conclusion, lower percentage of left ventricle systolic dysfunction in our DM1 cohort compared to some other studies may be caused by differences in patients' population, selection bias, and methodological issues. We must say although systolic dysfunction was reported in significantly higher percentage in some series of DM1 patients, meta-analysis showed an overall prevalence of 7.2%, which is similar to our results.[9]

Conflicts of interest

The authors declare that they have no conflicts of interest related to the subject matter or materials discussed in this article.

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