Author's Reply to Peverill: "Predictors of Left Ventricular Dysfunction in Friedreich's Ataxia in a 16-Year Observational Study"

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We appreciate the comments of Dr Peverill [1] regarding our 16-year observational study of adult patients with Friedreich ataxia (FA), a rare mitochondrial disease caused by guanine-adenine-adenine (GAA) repeat expansions in the *FXN* gene. As a decrease in the left ventricular ejection fraction (LVEF) is associated with a worse prognosis [2], our objective was the early identification of patients with a high risk of developing left ventricular systolic dysfunction. In particular, we wished to pinpoint the moment at which deterioration begins.

Based on our data, we built a model to predict a decrease in the LVEF below 50%, with a median of 5 years before it occurs. We showed that, aside from the genetic abnormality (length of shorter GAA repeats), echocardiographic follow-up may help to identify such patients on the basis of an increase in the left ventricular end-diastolic diameter (LVEDD) and septal wall thickness (SWT).

Normalization of the values for the size of the left ventricle by body surface area could have improved the predictive value of the model; however, the left ventricles of men are larger than those of women, even after normalization for body surface area, as demonstrated in healthy subjects [3]. Peverill et al. reported the same observations in FA patients [4]; thus, the simple normalization of left ventricular volume to body surface area would not have eliminated sex differences. Moreover, it would have been difficult to build a predictive model based on sex for this rare disease, for

which there were so few events over time (12%). We agree that these points should have been discussed in the paper, but we do not believe that this compromises the message, which is to regularly perform an echocardiographic examination of FA patients to detect early left ventricular abnormalities.

Compliance with Ethical Standards

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Conflicts of interest Francoise Pousset, Lise Legrand, Richard Isnard, and Alexandra Durr declare they have no potential conflicts of interest that might be relevant to the contents of this reply.

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