LETTERS TO THE EDITOR

The Effects of Tranilast on Cardiomyopathy in Becker Muscular Dystrophy Requires Profound Cardiac and Neurologic Evaluations

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To the Editor

We read with interest the article of Matsumura et al., who reported a pilot study of tranilast in two patients with dilated cardiomyopathy and muscular dystrophy (1). After therapy with 300 mg/day, the natriuretic peptide levels of the patients regressed. We have the following questions and concerns:

It remains unclear why the study was conducted for only three months, which is a rather short time for the observation of effects on heart failure therapy. Randomized studies about heart failure therapy generally have follow-up durations of at least two years (2). Furthermore, both patients received only 10 mg/d enalapril, which is lower than the dosage recommended under the current guidelines (3). What were the reasons for not up-titrating the enalapril dosage?

We did not notice any data on the patients' blood pressure. Tranilast has been reported to induce electrocardiography (ECG) changes comprising T-wave inversion, atrial fibrillation and ST-T wave changes (4, 5). Did ECG, performed four times during the study, show any new abnormalities or QT-prolongation? Did Holter ECG, performed three times during the study, show any arrhythmia?

The methods mention that blood tests, comprising the measurement of white blood cells and eosinophils, were performed. However, the results of these tests were not described. Did the patients develop eosinophilia, which is a side effect of tranilast (4, 5)?

Did the neurological situation change during the therapy with tranilast? Is the pharmacological effect of tranilast specific for cardiomyocytes, or does it also work in skeletal muscle? Concerning patient 1, it would be interesting to know the onset of the neuromuscular manifestations and if the progression of cardiac disease correlated with the progression of the neurological impairment. Concerning patient 2, the neurological diagnosis is unclear. What were the findings on the muscle biopsy? Did the patient show dystrophin deficiency, or did histology only reveal dystrophic features? Given that patient 2 had been on non-invasive ventilation (NIV) for 19 years, the muscular respiratory insufficiency does not appear to have been progressive. Did weakness and wasting of the limb muscles progress or remain stable during the 19 years on NIV?

Is the pharmacological effect of tranilast on cardiomyocytes specific for patients with muscular dystrophy, or does it also work in patients with dilated or ischemic cardiomyopathies?

The case report could benefit from a more detailed description of the clinical neurologic findings, the findings from the muscle biopsy, and the cardiac course during tranilast therapy. It would also be helpful to know if the family history of the two reported patients was positive for myopathy or cardiac disease.

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References


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