Letter to the Editor

HyperCKemia instead of Hyperkalemia in Chorea-Acanthocytosis

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Key Words

Chorea-acanthocytosis • HyperCKemia • Rhabdomyosarcoma cell line

To the Editors,

The group of Florian Lang is to be congratulated for their continued productive efforts to elucidate pathomechanisms of the neurodegenerative disease chorea-acanthocytosis (ChAc). Lang and collaborators have employed a variety of cell models to identify the functions of the ChAc protein, chorein, and most recently further specified its role for rhabdomyosarcoma cell survival in this journal [1].

With all respect, however, we must point out a continuing error in their series of publications. When the condition of ChAc is introduced, it is regularly described by these authors as being characterized by “chronic hyperkalemia” [1-7]. This notion can lead to erroneous assumptions in the clinic, in discussions of pathophysiology, and in experimental design or interpretation. Hyperkalemia has never been noted to be a clinical feature of ChAc over many years of clinical observations [8-9]. A dictation, spelling, or proofing error must have caused the mistake, and clearly “hyperCKemia” was originally intended. Elevated serum levels of creatine kinase (CK) are a well-recognized common finding in ChAc and have long been discussed as indicative of skeletal muscle cell membrane dysfunction. This observation might be of relevance for the ZF alveolar rhabdomyosarcoma cells that were studied by Yu et al. as they derive from skeletal muscle.

We also noted incidentally that reference 4 in their current bibliography appears irrelevant to the current work. We strongly suspect that the authors intended to refer to another work in which ChAc was discussed with respect to possible heart muscle involvement [10]. The erroneous reference to a study of sudden death in childhood cardiomyopathy by the same authors Mohiddin and Fananapazir [11] can be traced through previous publications [2,7]. We conclude that an initial mistake of author attribution became reiterated in the course of manuscript production.

For future publications, we would like to encourage Lang and colleagues to minimize confusion by ensuring that their errors are not propagated further.

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