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Cardiomyopathy in Celiac Disease: Carnitine Behind?

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Abstract In Volume 8, Issue 2 of the International Journal of Celiac Disease, Shah et al. presented a peculiar celiac disease presenting as cardiomyopathy in an adult female. We would highlight the importance of carnitine deficiency in such cases, both at diagnosis as well as during follow up; carnitine being widely reported as a cause of cardiomyopathy in celiac and non-celiac population, and supplementing these patients by carnitine might reverse this heart dysfunction.

Keywords: *celiac disease, cardiomyopathy, carnitine*

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1. Introduction

Among the interesting case reports recently published in the International Journal of Celiac Disease, Shah et al. presented a peculiar celiac disease (CD) presenting as a dilated cardiomyopathy in an Indian adult female [1].

We address the cornerstone role of carnitine deficiency in such cases, both at diagnosis as well as during follow up. Carnitine is more and more reported as a cause of cardiomyopathy in celiac and non-celiac population, and supplementing these patients by carnitine might reverse this heart dysfunction [2,3,4,5].

In this article, we give a glance of dilated cardiomyopathy due to carnitine deficiency in celiac patients.

2. Carnitine Deficiency and Cardio-myopathy

Carnitine is needed for the transfer of long-chain fatty acids across the inner mitochondrial membrane for subsequent β-oxidation. Carnitine can be synthesized by the body and is also obtained in the diet through the consumption of meat and dairy products [6].

Levo-carnitine is a natural and biologically active amino -acid derivatives and a micronutrient that plays a crucial role in human lipid metabolism and also in mitochondrial defence. It contributes to a myriad of physiological activities and has been proposed for treating an expanding range of cardiac problems including myocardial injury and cardiomyopathies [7,8].

It is well-known that cellular metabolism of fatty acids does require both intra-cytosol carnitine cycle and intra-mitochondrial β-oxidation cycle; and carnitine is primordial for translocation of long-chain acyl-CoA across the inner mitochondrial membrane; and thus plays a pivotal role in cardiac cells' contraction [9,10].

3. Carnitine and Cardiomyopathy in Celiac Population

Several cases of celiac patients presenting a dilated cardiomyopathy due to a secondary carnitine deficiency in CD related malabsorption are reported in the scientific literature, sometimes as a unique extra-intestinal manifestation [11-15].

Notably, most of CD associated cardiomyopathy cases are reported in childhood [16], probably as a result of the profound deficiency in the pediatric population**.**

For example, a monocentric Brazilian study found that CD prevalence in pediatric patients with dilated cardiomyopathy or myocarditis was as high as 1.8% [17].

Biologically, patients presenting a dilated cardiomyopathy with CD show a deeper decrease in serum carnitine levels than those presenting an isolated dilated cardiomyopathy and gluten-free diet leads to a progressive restoration of normal carnitine concentrations [18].

A recent Saudi investigational study reported a good correlation between the cardiac functions and the degree of intestinal mucosal injury and advocating for the use of Doppler tissue imaging (rather than conventional two-dimensional echocardiography) to detect early myocardial dysfunction in children with CD. [19]

A strict gluten withdrawal in these cases would restore a normal intestinal permeability and normalize the carnitine serum and then cellular levels, allowing a progressive reversal of the cardiomyopathy [20].

Of course, carnitine supplementation should be prescribed at first-line if a clear deficiency is documented

or even in highly suspected cases (if carnitine dosage is not available).

In sum, a dilated cardiomyopathy due to carnitine deficiency may occur in CD patients and carnitine deficiency may present not only at the time of diagnosis of the CD but it may also develop during the gluten-free diet, particularly in patients with fast weight gain and without carnitine supplementation. [21].

4. Conclusion

Co-occurrence of dilated cardiomyopathy and CD should emphasize the prompt diagnosis of carnitine deficiency, even without systemic symptoms (digestive manifestations of CD and/or muscular signs of carnitine deficiency). Strict, long-life gluten-free diet with adapted carnitine supplementation reverse this heart dysfunction. Close management and follow up are mandatory.

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