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Prevention o. Diabetic Cardiomyopathy in Pediatrics ?

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Background: The development of Diabetic cardiomyopathy (DCM) is multifactorial and several pathophysiologic mechanisms have been proposed to explain structural and functional changes associated with DCM. α-Lipoic acid (ALA) . powerful antioxidant may has. protective role in diabetic cardiac dysfunction.

Aim of the work: This study aimed to assess the potential role of oxidative stress, inflammatory cytokines, apoptosis and fibrosis in diabetic cardiac insult. It also investigated the possible protective role of α-lipoic acid on diabetic left ventricular (LV) dysfunction in type. diabetic children and adolescents.

Subjects and methods: 30 patients were randomized to receive insulin treatment (n. 15) or insulin plus α-lipoic acid 300 mg twice daily (n. 15). Age and sex matched healthy control children and adolescents (n. 15)were also included. Patients were evaluated with conventional2-dimensional echocardiographic examination (2D), pulsed tissue Doppler (PTD), and 2-dimensional longitudinalstrain echocardiography (2DS) before and after therapy.3D strain (longitudinal,circumferential,area, and radial strain) were estimated. Plasma level of glutathione, malondialdhyde (MDA), nitric oxide, tumor necrosis factor-α (TNF-α), Fas Ligand (Fas-L), matrix metalloproteinase-2 (MMP-2) and troponin-I were determined before and after treatment.

Results: Diabetic patients had significant lower level of glutathione and significant higher levels of malondialdhyde (MDA), nitric oxide, tumor necrosis factor-α (TNF-α), Fas Ligand (Fas-L), matrix metalloproteinase-2 (MMP-2) and troponin-I than control subjects. Increased expression of transforming growth factor-β (TGF-β) mRNA in peripheral blood mononuclear cells was also observed in diabetic patients. 2D global longitudinal strain and 3D longitudinal, circumferential and area strain were significantly decreased in diabetic children. α-lipoic acid significantly increased glutathione level and significantly decreased MDA, nitric oxide, TNF-α, Fas L, MMP-2 ,troponin. levels and TGF-β gene expression levels. Moreover, α-lipoic acid significantly increased mitral e/a ratio, ventricular global peak systolic strain in diabetic patients. There were significant negative correlation between Global peak systolic strain (G) and glutathione and significant positive correlations between. and MDA ,NO. TNF-α. and Fas-L. In addition,. significant positive correlation between e/a ratio and glutathione (r. 0.515) and significant negative correlations between e/a and MDA ,NO. TNF-α. and Fas-L were also observed

Conclusion: These data suggest that oxidative stress, inflammatory cytokines such as TNF-α, apoptosis and fibrosis play. role in the development of diabetic cardiac dysfunction and that α-lipoic acid may have. beneficial role in the management of type. diabetic patients as cardioprotective therapy and prevention of development of of diabetic cardiomyopathy.

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Woodhouse–Sakati syndrome:. common syndromic form of diabetes among arabs

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Hypogonadism, alopecia, diabetes mellitus, mental retardation, and extrapyramidal syndrome [also known as Woodhouse–Sakati syndrome (WSS)] is rare autosomal recessive neuroendocrine and ectodermal disorder. The diagnosis is confirmed in. proband with suggestive clinical findings and biallelic pathogenic variants in DCAF17 (formerly known as C2orf37) on molecular genetic testing. Diabetes mellitus (DM), either type. (insulin-dependent) or type. (non-insulin-dependent), was reported in 62% of individuals under age 25 years and 95% of those over age 35 years. To date 32 families (76 affected individuals) have been reported. We report on an additional 20 patients from. highly consanguineous Tribe from Qatar, presenting with interfamilial phenotypic variability. We will describe in details the natural course and the interfamilial phenotypic variability of WSS that may lead to challenges in making the diagnosis. In addition, to alert Endocrinologist to this important syndromic form of DM and highlights that WSS probably not as rare among the Arabs as previously thought.

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