

8. DILATED CARDIOMYOPATHY IN ASSOCIATION WITH THE ALCAPA SYNDROME

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Introduction: Dilated cardiomyopathy is a severe condition in which the heart muscle is weakened and no longer has the strength to pump blood throughout the body. The weakened heart is unable to pump more blood, therefore more blood remains to this level after each heartbeat. As larger amounts of blood remain in the lower chambers of the heart, they expand. Over time, the heart muscle loses its shape and becomes increasingly weaker.

Objectives: Study of clinical evolution and ECG of patients diagnosed with associated dilated cardiomyopathy or ALCAPA Syndrome from a basic treatment with IEC, diuretics and B-blocking agents along with the surgical treatment.

Materials and Methods: 10 children of which 9 diagnosed with dilated cardiomyopathy and 1 child who associates ALCAPA syndrome were evaluated by Ross score, NYHA and echocardiographic. Cardiac index measurement using ECG and CT after receiving basic treatment.

Results: The 9 patients with dilated cardiomyopathy treated by the basic therapeutic method have evolved to improve cardiac indexes (Ross and NYHA) showing an increase in left ventricular ejection fraction gather up 20%. The patient with dilated cardiomyopathy and ALCAPA syndrome showed no improvement following therapy being directed to surgical treatment.

Conclusions: Dilated cardiomyopathies in pediatric age have an 80% response rate to basic treatment with IEC, diuretics and B-blockers that lead to Ross and NYHA amelioration. Dilated cardiomyopathy associated with ALCAPA syndrome has one treatment option: heart transplant.

Keywords: evolution, dilated cardiomyopathy, treatment, children, ALCAPA syndrome

9. CLINICAL CHARACTERISTICS OF GRAVES' ORBITOPATHY

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Introduction: Graves' orbitopathy (GO) constitutes a major clinical and therapeutic challenge. GO is an autoimmune disorder representing the most common and most important extrathyroidal manifestation of Graves' disease. Although the pathogenesis of GO is beyond the scope of this study, attention is drawn to the link between the orbit and thyroid, which has important clinical and therapeutic implications. Optimal management of GO requires a coordinated approach addressing the thyroid dysfunction and the orbitopathy.

Purpose and Objectives: To establish the relationship between Graves' disease evolution and the onset of ocular manifestations. To evaluate the activity and severity of GO in our study group.

Materials and Methods: This study is based on the clinical examination of 16 patients, using the Clinical Activity Score (CAS) and the NOSPECS classification of the disease's severity.

Results: During this study, we have seen 16 patients (32 eyes). 12 of these patients presented bilateral involvement, while 4 – unilateral involvement. We have studied the onset of Graves' orbitopathy in relation to the onset of hyperthyroidism, and we've determined that most commonly GO occurs at the same time or follows the hyperthyroidism (up to 81% of cases). The activity of GO was determined using CAS. We've determined that the spontaneous orbital pain, gaze evoked orbital pain, conjunctival redness and the eyelid swelling were the most common manifestations presented by our patients (50–75% of cases), while eyelid erythema was the less common (12.5%). The frequency of active and inactive cases was almost equal, with a minor prevalence of active processes. The disease's severity was appreciated using NOSPECS classification. Most patients presented a minimal grade of severity (43.7% – 7 patients), while 37.5% (6 patients) have presented a moderate grade and only 18.8% (3 patients) – maximal severity. The first two classes of the