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[Cardiac involvement in metabolic diseases]

[Article in Portuguese]

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Abstract

OBJECTIVE: To evaluate cardiac involvement in children with metabolic disease in the out patient clinic of the Pediatric Cardiology Unit of Maria Pia Children's Hospital and their follow-up.

MATERIAL AND METHODS: Twenty-nine medical records belonging to out patients with metabolic disease in consultation at our unit were reviewed. The following data from each record was analyzed: sex, metabolic disease diagnosis, age and motive for referral to a pediatric cardiology unit, cardiology diagnosis, therapy and evolution.

RESULTS: Seventeen patients were boys and 12 girls. The average age of referral was 7.2 years (SD 4.8). The motives for referral were: screening for heart disease, 16; heart murmur, 7; congestive heart failure, 3; heart murmur and fatigue, 2; poor weight gain, 1. The following metabolic diagnoses were made: lysosomal diseases, 21; mitochondrial citopathies, 5; disorder of beta-oxidation of fatty acids, 2; carbohydrate deficient glycoprotein syndrome (CDG syndrome), 1. The cardiologic evaluation was normal in ten patients (4 with lysosomal disease, 4 with mitochondrial citopathy, one disorder of beta-oxidation of fatty acids, the CDG syndrome). Mitral and aortic valve lesions predominated in lysosomal diseases (12/21); myocardial involvement alone was present in two patients, and both myocardial and valvular lesions were present in three. Dilated cardiomyopathy was the presented manifestation in two patients-one with mitochondrial citopathy and one with a disorder of beta-oxidation of fatty acids. Three patients died and 26 remain out-patients. One patient was submitted to valve surgery. The average duration of follow-up was 21 months (SD 24).

COMMENTS: Lysosomal diseases were the most representative in our patients, as described in the literature. Heart valve disease was the most frequent alteration. Indication for heart valve surgery is

dependent on systemic involvement of the primary disease. All children with a metabolic disease with eventual heart involvement should be evaluated periodically by a cardiology unit. On the other hand, it is mandatory to screen a cardiomyopathy of unknown cause for a metabolic disease. The authors draw attention to the importance of infectious endocarditis prophylaxis in this group of patients.

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