

### **Epidemiologic Data, Prognosis and Biopsy Guided Therapy in Children with Myocarditis and Cardiomyopathy**

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**Introduction:** Myocarditis and cardiomyopathies are acquired causes of heart failure with a wide range of aetiology, pathophysiology and prognosis. In Greece there is a paucity of data regarding this group of diseases in the paediatric population. This study reports epidemiologic, management and prognosis data of children with myocarditis and cardiomyopathy, referred to a tertiary center because of cardiac failure.

**Methods:** We performed retrospective data collection for paediatric patients (0-18 years old) diagnosed with myocarditis / cardiomyopathy and hospitalised from 2004 to 2017. Analysis of data was performed on the basis of demographics, diagnosis, biopsy results, management and prognosis.

**Results:** 80 patients were analysed (44 boys; 36 girls), with the following diagnoses: 37 cardiomyopathy (26 dilated; 4 hypertrophic; 5 restrictive); 2 arrhythmogenic right ventricular dysplasia and 43 myocarditis. We defined the following age groups for age distribution: 0-1 year old (n=13); 1-5 y.o. (n=25); 5-16 y.o. (n=37) and >16 y.o. (n=5). Diagnosis was based on echocardiography findings, myocardial enzyme levels and biopsy findings [active inflammation; necrosis; hypertrophy; fibrosis; (near) normal; other]. All patients but 3 underwent cardiac biopsy. Overall, 27 cardiac histological specimens (23 out of the 'myocarditis' group and 4 out of the rest) were positive for viral or bacterial genetic material through polymerase chain reaction (PCR) testing. Of the positive patients, 21 received targeted therapy (antiviral or antibiotic agents). No specific therapy was available for the remaining patients at the time of diagnosis. Based both on biopsy findings (active inflammation or not) and on PCR in blood/cardiac specimen, 20 patients in total received intravenous immunoglobulin and 4 systemic corticosteroids. During follow up, 11 children (15 %) died; 9 (11%) were transplanted or are in the transplant list; 9 (11%) required prolonged medical support for heart failure; the rest (63%) improved or had full recovery.

**Conclusions:** Acquired heart failure in the paediatric population is mostly due to myocarditis and dilated cardiomyopathy, while the majority of affected patients survive over long follow up. Cardiac biopsy and PCR testing in blood and cardiac specimen are very useful tools for diagnosis as well as in guiding specific treatment, especially in patients with myocarditis.