

# EPIDEMIOLOGIC DATA, PROGNOSIS AND BIOPSY GUIDED THERAPY IN CHILDREN WITH MYOCARDITIS AND CARDIOMYOPATHY

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## INTRODUCTION:

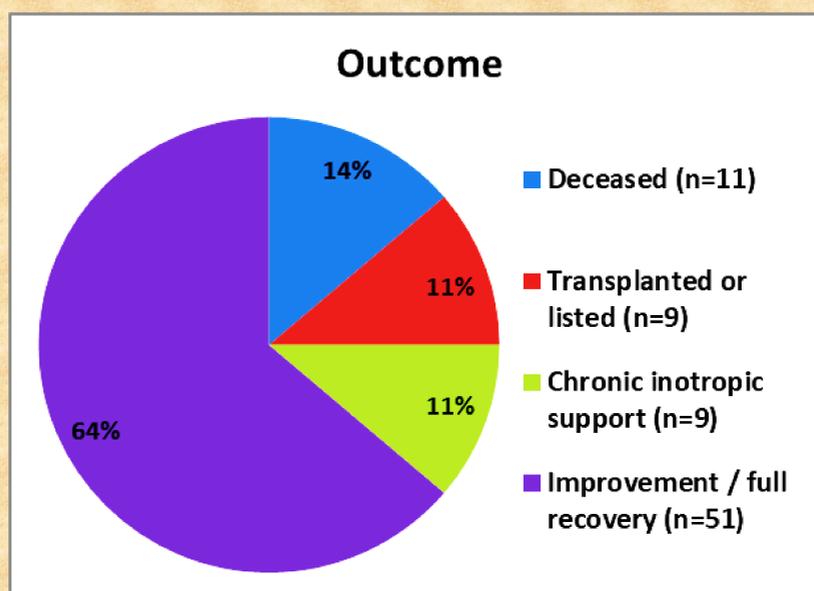
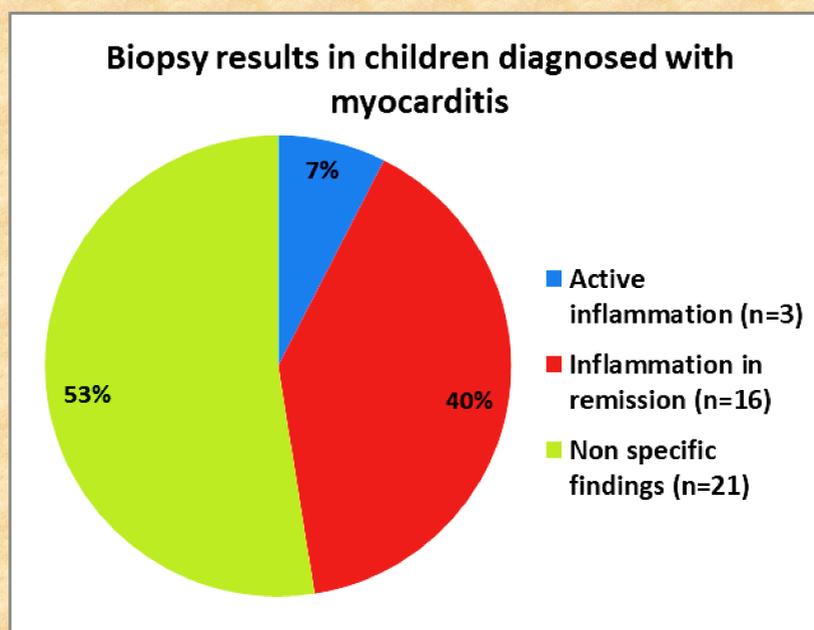
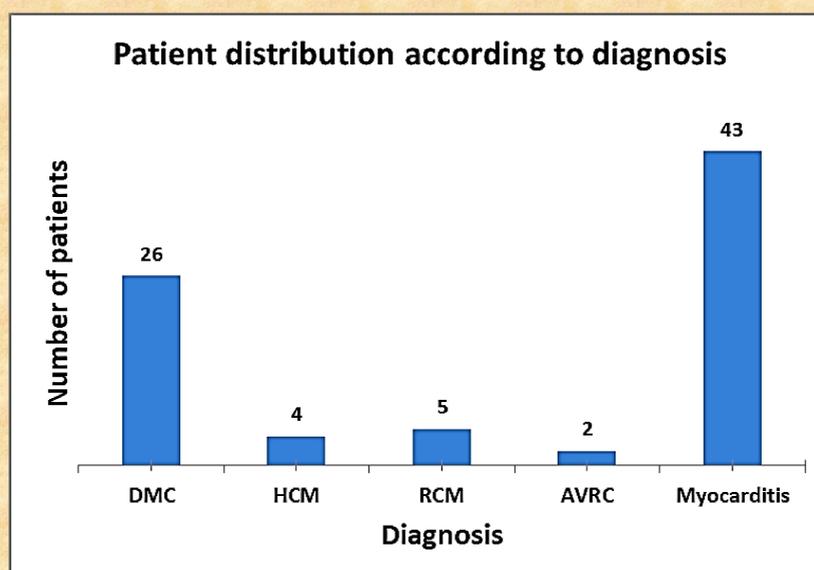
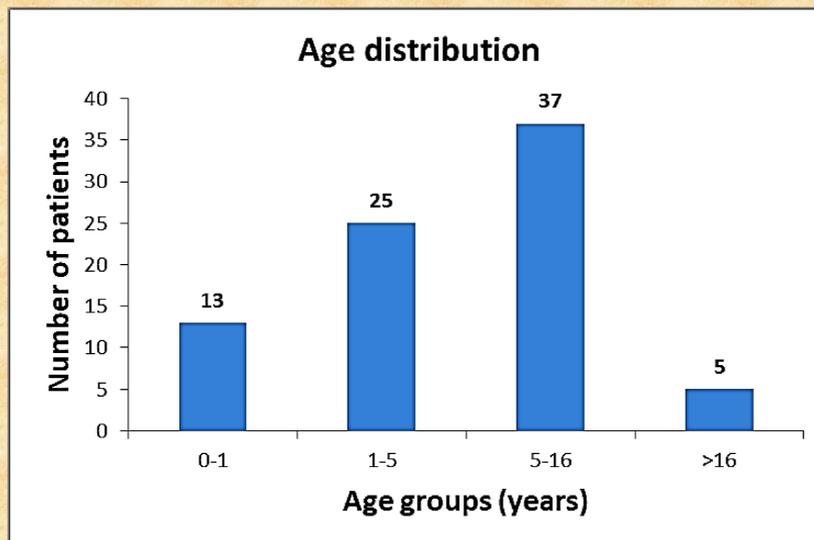
- Myocarditis and cardiomyopathies are acquired causes of heart failure with a wide range of aetiology, pathophysiology and prognosis.
- In Greece there is a paucity of data regarding this group of diseases in the paediatric population. This study reports epidemiologic, management and prognosis data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

## METHODS:

- We performed retrospective data collection for paediatric patients diagnosed with myocarditis / cardiomyopathy and hospitalised from 2004 to 2017. Age ranged from 1 day to 18 years old.
- Analysis of data was performed on the basis of demographics, diagnosis, biopsy results, management and prognosis.
- The smallest patient weighed 3 kgs.
- Diagnosis was based on cardiac echo findings, myocardial enzyme levels and results from myocardial biopsy [active inflammation; necrosis; hypertrophy; fibrosis; (near) normal; other].

## RESULTS:

- 80 patients were analysed (44 boys; 36 girls). We defined groups for age distribution as depicted.
- 37/80 patients (46%) were diagnosed with cardiomyopathy (26 dilated; 4 hypertrophic; 5 restrictive); 2 with arrhythmogenic right ventricular dysplasia and 43/80 (54%) with myocarditis.
- 40/80 patients (50%) were hospitalised in the Paediatric Intensive Care Unit. 3 patients (3.7%) were transferred intubated to our center. 17/80 patients (21%) were already on IV inotropic support on admission.



## RESULTS:

- Median value for NT-proBNP for the population studied was 9248 pg/ml (95% CI 7611-16930).
- All patients but 3 (because of severe haemodynamic instability) 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.
- Out of the positive patients, 21 received targeted therapy (antiviral or antibiotic agents). No specific therapy was available for the remaining 6 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.
- Our findings are in accordance with published literature regarding aetiology of heart failure in paediatric patients with anatomically intact heart.
- 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.