

## Baseline features and management in adult and pediatric clinically suspected and biopsy-proven myocarditis in the cardiomyopathy and myocarditis long-term EORP registry

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**Background:** The Myocarditis section of the EORP Cardiomyopathy and Myocarditis Long-term Registry is a prospective, observational, multinational registry of adult and pediatric patients enrolled using the ESC 2013 diagnostic criteria of clinically suspected (CS) or biopsy-proven (BP) myocarditis (myoc).

**Purpose:** i) To obtain a real-world snapshot of features and management of myoc; ii) to assess features at presentation in CS and in BP myoc and by age.

**Methods:** 581 patients (68% male), 493 adults, aged 34.9 (SD 18.5) years, and 88 children, aged 8.1 (SD 5.2) years, were divided into 3 groups (G): G1 (n=234, 40%), CS myoc plus cardiac magnetic resonance (CMR) confirmed; G2 (n=222, 38.2%), BP myoc; G3 (n=125, 21.5%), CS myoc, no or normal or inconclusive CMR. Baseline features, procedures, medications were analysed in the total population, in adults vs children, and among G.

**Results:** In all patients: pseudo-infarct presentation with normal coronary arteries is common (58%), as is heart failure (HF) with or without chest pain and troponin release (58%), followed by arrhythmia (41.9%). In children new-onset HF is more common than in adults (29/32, 90% vs 90/190, 47%, p=0.001). In both adult and pediatric G2 BP myoc, HF and arrhythmia were more common than in CS myoc. Left and right ventricular (RV) echocardiography and CMR function indexes and troponins were lower, NT-pro BNP was higher in G2 BP myoc vs G1 and G3 CS myoc. On

CMR oedema and/or Late Gadolinium Enhancement (LGE) were found in 57.4% of adult and in 31.3% of paediatric G2 BP myoc. Endomyocardial biopsy (EMB) was obtained in a similar proportion in children (31/88, 35.2%) and adults (185/493, 37.5%, p=NS), ventricular assist devices were more commonly implanted in G2 children (8/32, 25%) than in G2 adults (4/190, 2.1%, p=0.001), ICD tended to be less common in G2 children (2/32, 6.3%) than in G2 adults (48/190, 25%, p=0.07). In all patients EMB, mainly RV (75.8%), had a low complication rate (4.7%), similar in adults vs children, with no procedure-related death. Histology findings were: lymphocytic myoc (78.9%), giant cell (10.9%), sarcoid (6.9%), non specific (16%). Viral genome was found in 44% of patients (most common PVB19, 21.7%, HHV6, 9.5%). In all patients HF and antiarrhythmic drugs were more frequently used in G2, antivirals in a patient minority, steroids in 24.7%, immunosuppression (IS) in 22.6%. In children steroids or IS were given regardless of G, in adults mainly to G2 BP myoc patients, in keeping with the ESC 2013 expert reco's.

**Conclusions:** EMB is safe in children and adults and is still the diagnostic gold standard, since CMR failed to identify myoc in a high proportion of G2 BP patients. Etiology-directed therapy was used in a minority of G2 cases, and/or regardless of etiology, thus there is room for improved management. G2 BP patients were older, sicker, had worse biventricular function, more medications and ICDs; follow-up may show their worse outcome.