Survival and outcome in pediatric myocarditis: Data of the German multicenter registry “MYKKE”


Deutsches Herzzentrum Berlin, Pediatric Cardiology, Berlin, Germany (1); Charité - Universitätsmedizin Berlin, Institute for Computational and Cardiovascular Medicine, Germany (2); DZHK (German Centre for Cardiovascular Research), partner site Berlin, Germany (3); Charité - Universitätsmedizin Berlin, Pediatric Cardiology, Berlin, Germany (4); Herzcentrum Leipzig, Pediatric Cardiology, Leipzig, Germany (5); Department of Pediatric Cardiology and Intensive Care Medicine, Hannover Medical School, Hannover, Germany (6); Universitätsklinikum Erlangen, Pediatric Cardiology, Erlangen, Germany (7); Universitäres Herzcentrum Hamburg, Pediatric Cardiology, Hamburg, Germany (8); Herz- und Diabeteszentrum NRW, Pediatric Cardiology, Bad Oeynhausen, Germany (9); Universitätsklinikum des Saarlandes, Pediatric Cardiology, Homburg, Germany (10); Klinikum Links der Weser, Pediatric Cardiology, Bremen, Germany (11); Universitäts-Herzzentrum Freiburg Bad Krozingen, Pediatric Cardiology, Freiburg, Germany (12); Universitätsklinikum Münster, Pediatric Cardiology, Münster, Germany (13); Universitätsklinikum Tübingen, Pediatric Cardiology, Tübingen, Germany (14); Deutsches Herzzentrum München, Technische Universität München, Pediatric Cardiology, München, Germany (15); Uniklinik Köln, Pediatric Cardiology, Köln, Germany (16); Klinikum der Universität München, Pediatric Cardiology, München, Germany (17); Universitätsklinik Gießen, Pediatric Cardiology, Giessen, Germany (18); Universitätsklinikum Ulm, Pediatric Cardiology, Ulm, Germany (19); Universitätsklinikum Bonn, Pediatric Cardiology, Bonn, Germany (20); Universitätsmedizin Göttingen, Kinderherzklinik Göttingen, Pediatric Cardiology, Göttingen, Germany (21); Universitätsklinikum Schleswig-Holstein, Pediatric Cardiology, Kiel, Germany (22); Kompetenznetz Angeborene Herzfehler, Berlin, Germany (23); Deutsches Herzzentrum Berlin, Cardiology, Berlin, Germany (24)

Objectives
Myocarditis is a major cause of severe heart failure in childhood. “MYKKE”, a multicenter prospective registry for pediatric patients with suspected myocarditis, aims with its ongoing enrollment to gain knowledge on incidence, diagnosis, therapy and outcome.

Methods
Since 2013 twenty centers actively included patients. A sub-analysis on clinical data and a multivariate analysis (MANOVA) of 195 patients enrolled until 2016 was performed according to baseline data and their impact on survival. Patients with typical findings in cardiovascular magnetic resonance (CMR) and/or endomyocardial biopsy (EMB) were defined as myocarditis positive.

Results
Until October 2017, 280 patients were enrolled, median (range) age 13.0 (0-17) years; 66.8% male. Three age groups were defined according to clinical course and severity of disease: 0-<2 (24.3%), 2-12 (20.7%) and 13-18 years (55.0%). 11% were listed for heart transplantation (HTx); and 6.8% subsequently underwent HTx. The overall mortality was 7.5%. Patients <2 years had a higher incidence (59%) of a severely reduced left ventricular ejection fraction (EF <30%) compared with age groups 2-12y (38%) and 13-18y (7%); p<0.001. They also had the highest rate of mechanical circulatory support (MCS; p=0.001) and death (p=0.003). A sub-analysis revealed that in 78% (n=153/195), EMB (n=44), CMR (n=40) or both (n=69) were performed with a positive result for myocarditis in 69%. Histology showed: 15% acute, 43% subacute/chronic, 12% status post, 1% eosinophilic or granulocytic myocarditis, 5% no infection, 3% DCM, 1% HCM, 7% other. The mortality in the MCS cohort was 21% (n=6/28). The proof of myocarditis had no impact on the possibility of weaning from MCS (p=0.675). Non-MCS patients showed a significantly better survival (97% vs. 75%; p=0.001). The MANOVA highlighted age (p=0.028) and EF (p=0.05) as dependent variables in predicting survival. Accordingly, patients <2 years and EF <30% carried the highest risk for mortality according to a fulminant course (p<0.001).

Conclusions
Our data underline that myocarditis stays a life-threatening disease in the pediatric population, especially in very young children. This first prospective data analysis enabled the definition of a high-risk group. These results emphasize the utility of “MYKKE” as a registry for suspected myocarditis in children and adolescents.