MP5-5

The MFS-HARM score: prediction of cardiomyopathic features of Marfan syndrome with the MarFan-Syndrom-Heart-At-Risk-Model

Diaz-Gil D.(1), Groth M.(2), Kodolitsch Y.V.(3), Kozlik-Feldmann R.(1), Lam H.V.(4), Mueller G.(1), Olfe J.(1), Rueffer A.(1), Stark V.(1), Tahir E.(4), and Mir T.S.(1)
University Heart Center Hamburg, Department of Pediatric Cardiology, Hamburg, Germany (1); University Medical Center Hamburg-Eppendorf, Department of Pediatric Radiology, Hamburg, Germany (2); University Medical Center Hamburg-Eppendorf, Department of General and Interventional Cardiology, Hamburg, Germany (3); University Medical Center Hamburg-Eppendorf, Department of Diagnostic and Interventional Radiology and Nuclear Medicine, Hamburg, Germany (4)

Background: Marfan syndrome (MFS) is a connective tissue disease frequently involving the cardiovascular system. Even though secondary cardiac manifestation in the form of valvulopathy is common, the question of primary cardiomyopathy still remains in debate. The purpose of this study is to understand the factors that predict cardiomyopathic features in pediatric MFS-patients.

Methods: Using data from 40 randomly selected MFS-patients from our pediatric Marfan database, echocardiography, and chart review, we developed a score predicting congestive heart failure (CHF) in patients with MFS. CHF was defined as either a cardiac index (CI) below the age adjusted lower limit of normal (LLNaa), a reduced ejection fraction (EF) below 50%, or the presence of clinical symptoms of CHF (NYHA >1). Using multivariable logistic regression analysis, predictors of CHF were determined and a score was postulated.

Results: In the 40 patients with secured MFS (age, mean±SD, 15.5±3.62 years) the CHF-incidence was 50%. Only 17.5% experienced symptoms of CHF (NYHA >1). The CI and the EF was on average lower in the CHF-cohort (mean±SD, CI 2.15±0.77 L/min/m2 vs. 3.03±1.29 L/min/m2, p=0.044; EF 60.62±12.81% vs. 67.72±11.76%, p=0.109). The presence of aortic valve regurgitation (22.5%) did not differ between the groups (p=0.268). Predictors of CHF were myopia >3 dpt, female sex, striae distensae, height <90th percentile, age <17 years, heart rate <LLNaa, and main pulmonary artery to sinus valsalva ratio > 0.7. Each predictor was assigned a point value of 1. The 7-point MarFan-Syndrom-Heart-At-Risk-Model (MFS-HARM) yielded a calculated area under the curve of 0.93. Each point increment was associated with a 8.27-fold increase in the odds for CHF (95% confidence interval, 2.44–28.00, p=0.001). An MFS-HARM score ≥3 predicts CHF with a 90% sensitivity and an 85% specificity.

Conclusion: In a randomly selected cohort of pediatric MFS-patients, 50% experience signs of CHF. The MFS-HARM, a composite score derived from patient characteristics tracked on follow-ups in a Marfan Center, predicts those at increased risk for CHF. After validation, risk prediction along a continuum of clinical and imaging parameters may open opportunities to focus clinical resources on the prevention of the of cardiac function deterioration in adolescents with MFS.