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N-BNP levels in pediatric Marfan patients - a predictor for left-ventricular pathologies of Ghent Criteria?

Stark V.C., Diaz-Gil D., Kozlik-Feldmann R., Mueller G.C., Olfe J., Mir T.S.
Clinic for Pediatric Cardiology, University Heart Center, Hamburg, Germany

Objective: Correlation of elevated N-BNP levels with reduced diastolic function as well as with dilatation of sinus of Valsalva (SV) was shown before in a collective of pediatric and adult Marfan patients. Isolated data for pediatric Marfan patients is missing and dynamic pathologies of the mitral valve(MV) and its correlation with N-BNP have not been investigated yet. We hereby evaluate correlation of SV dilatation, aortic valve regurgitation(AVR) and MV Marfan pathologies with N-BNP in an isolated pediatric collective. To individualize follow-up regime and therapy our pediatric patients would benefit from early risk stratification.

Methods: Between 2008 and 2018, we diagnosed Marfan syndrome in 171 patients (9.3±5.4y;m:92(54%)) in our specialized pediatric Marfan clinic. In routine follow-up visits, we examined children according to RGC including echocardiography and measured N-BNP with every blood sample. We analyzed N-BNP with age-related standard values 146 times and correlated N-BNP with left ventricular Marfan pathologies (SVdilatation, AVR, MVprolaps, MVregurgitation). In addition, we analyzed N-BNP in 82 patients with medication in comparison to 64 patients without.

Results: N-BNP was not elevated in absence of left ventricular pathologies. Patients with SVdilatation developed higher N-BNP (262±656ng/l) than those without (70±111ng/l;$p<0.01$). AR patients developed higher N-BNP (376±831ng/l) than patients with competent AV (89±135ng/l,$p<0.01$). Patients receiving a combined treatment with Sartan and Betablocker(BB) showed significantly higher N-BNP (644±1294ng/l) than patients without treatment (120±79ng/l;$p<0.01$). In patients treated with a Sartan or BB alone N-BNP levels did not differ significantly in comparison to patients without treatment. MV pathologies did not correlate with higher N-BNP.

Conclusion: As soon as the SV dilates or the aortic valve regurgitates, pediatric Marfan patients´ N-BNP is significantly higher than of those without. But MV pathologies were not associated with N-BNP elevation in our pediatric collective. Thus, unfortunately, N-BNP levels do not gain additional information in pediatric Marfan patients with normal diastolic and systolic function. Nevertheless, it may compliment and affirm existing pathologies like SVdilatation. In conclusion, measurement of N-BNP supplements care of pediatric Marfan patients but regular clinical follow-up examinations including echocardiography and MRI are indispensable and remain gold standard for a safe patient care.