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Von Willebrand Factor parameters, biomarkers for disease activity and Coronary artery lesion in Kawasaki disease

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Objectives: Von Willebrand factor (VWF), a large multimeric glycoprotein is essential for primary haemostasis. Increased plasma levels of VWF:Antigen (VWF:Ag) have been observed in cardiovascular diseases or vasculitis and are presumably caused by activation of the endothelium. The impact of Kawasaki disease (KD), a vascular inflammatory disease, regarding VWF:Ag, VWF:Collagen binding activity (VWF:CB) and VWF multimers structure analysis, has not been clarified yet. We investigated VWF-parameters in patients with Kawasaki disease (KD) as a surrogate marker for disease activity and Coronary artery lesion (CAL).

Methods: 28 KD patients, 10 with Coronary artery lesion were enrolled to this study. In 5 patients serial measurements were collected. VWF:Ag and VWF:CBA were determined by enzyme-linked immunoassay. The VWF:CB/VWF:Ag -ratio was calculated and the VWF structural features were assessed by multimer structure analysis. We evaluated the correlation between VWF parameters and standard inflammatory markers. The impact of the patients´ age, point of time of blood collection, being refractive to therapy and CAL on the VWF parameters was assessed. We furthermore analyzed the VWF-parameters predictive value for CAL.

Results: VWF:Ag and VWF:CB levels were significantly higher in the acute phase as compared to the convalescence phase. There was a moderate positive correlation (Pearson coefficient > 0,3) of all VWF parameters with CRP and VWF:Ag with a high platelet count. A lower VWF:CB/VWF:Ag-ratio was negatively correlated with a higher leucocyte count. Interestingly, the VWF:CB/VWF:Ag -ratio was significantly decreased in those patients with CAL (mean 0.96 vs. 0.64; p= 0.036) whereas the absolute levels of VWF:Ag and VWF:CBA did not show any differences with respect to CAE/CAA. Using a model to predict CAL, the AUC of the ROC was 0,84 (sensitivity of 60% and specificity of 94%). Those patients with very low VWF:CB/VWF:Ag-ratio in the acute phase had persistent CAL (1 year follow up).

Conclusion: Our study indicates that a comprehensive analysis of VWF-parameters may help to monitor KD inflammation and furthermore may help to detect those patients with increased risk for CAL. Further analyses should be performed in a larger study population.