

Abnormal coagulation factors in single ventricle physiology patients: correlation with hemodynamic parameters and its impact on the postoperative course

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Introduction: Thromboembolic events significantly influence morbidity of patients with univentricular hearts. The presence of an intrinsic or acquired hypercoagulable state has been considered before in these patients. This study evaluates coagulation profiles in a cohort from Norwood stage I through Fontan palliation (stage III) in relationship to liver function, hemodynamic variables and clinical outcome.

Methods: Twenty-six consecutive patients with single ventricle anatomy were included between 2016-2018 and blood samples for assessment of an advanced coagulation profile were taken during cardiac catheterization. Coagulation parameters and invasive preoperative hemodynamic parameters, as well as the clinical course from birth to six months after sampling were retrospectively analyzed.

Results: Mean (\pm SD) age and weight at time of blood sampling was 76 ± 20 months and 10 ± 4.5 kg, respectively. Ten patients were included before stage II, 13 before and 3 after stage III. Ventricular anatomy was left dominant in 17 patients. Sixteen patients (16/26; 62%) showed decreased antithrombin (AT) and/or protein-C (PC) and/or free-protein-S (PS)-function and/or free-PS-antigen. Two patients showed abnormal activated-protein-C-resistance-ratio due to heterozygous factor-V-Leiden mutation and one heterozygous prothrombin G20210A mutation. Group comparison [abnormal coagulation profile (group 1; n=16) versus normal coagulation profile (group 2; n=10)] showed longer postoperative hospitalization time ($p=0.04$), longer postoperative catecholamine support ($p=0.01$), a higher incidence of thromboembolic events ($p=0.04$) and chylothorax ($p=0.007$) in group 1. In 5/16 (31%) group 1 patients thromboembolic complications occurred: cerebral stroke (n=1), intestinal ischemia (n=2), thrombus formation in inferior caval vein (n=1) and pulmonary vein (n=1). Extensive, prolonged bleeding was seen in two patients.

Mean pulmonary artery pressure negatively correlated with PC ($p=0.02$), free-PS-function ($p=0.02$), and free-PS-antigen ($p=0.003$). Pulmonary wedge pressure negatively correlated with PC ($p=0.03$). Oxygen saturation correlated positively ($p=0.04$) and pulmonary resistance (R_p) negatively ($p=0.01$) with AT. No correlation was found for anthropometric parameters, stage of palliation, ventricular morphology, end diastolic systemic ventricle pressure and transpulmonary gradient, prothrombin and activated partial thromboplastin time, fibrinogen, liver transaminases, hemoglobin, and platelet count.

Conclusions: Abnormalities in coagulation parameters are common in patients with single ventricle physiology. There is a correlation between hemodynamic and coagulation parameters. Coagulation abnormalities negatively affect postoperative course.