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The first treatment experience of a patient with functional single ventricle and homeostasis pathology

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Introduction: Homeostasis pathology increases significantly risk of death after CHDs correction. We observed 5 cases of development of venous thrombosis in patients with “functional single ventricle” after BCPC and TCPC. We assessed the methylenetetrahydrofolate reductase thermolabile variant (MTHFR C677T), the prothrombin mutation (FII G20210A), the factor V mutation (FV A4070G and G1691A), and the platelet glycoprotein receptor IIIA (GpIIIa PlA1/A2) as predictors of thrombosis.
Results: We had a 6-year-old patient with functional single ventricle, heterotaxy with polysplenia syndrome, absence of subhepatic segment of inferior vena cava, arteriovenous malformation of pulmonary vessels and heterozygous carrier of allele PL-A2 of GpIIIa gene, presence of which determines increase of thrombocyte aggregation and resistance to antiaggregant therapy. At 1 year old Kawashima procedure, main pulmonary artery ligation were performed. At examination the patient’s weight was 19 kilos, saturation– 53%-59%, antithrombin deficiency III – 73% (norm – 80-140%), protein C decrease - 67% (norm – 10-130%), level increase of soluble fibrin-monomeric complexes to 7,5mg% (norm – to 3,5%), erythrocytes level to 6.99*109 /l and Hb to 17,7 g/dL, hematocrit 57%. Performed surgery: redirection of hepatic venous into v. azygous by prothsesis «Gore-tex» 16 мм, left pulmonary artery banding, atrioventricular valve repair. Postoperative monitoring of homeostasis system included: determination of antitrombin III, activated partial thromboplastin time, D-dimer, INR, level of thrombocytes and hematocrit. Level of antithrombin III was defined daily during 9 days. When the level was lower, intravenous infusion of antithrombin III human was prescribed till target level 90% with simultaneous infusion of fresh frozen plazma in dose 10 ml/kl/daily. In 6 hours after surgery heparine infusion in dose 5 un/kg/hour with further dose increase to15 un/kg/hour was started. On 5th day after surgery heparine was canceled, fraxiparine was prescribed 0,3 ml twice per day. From 6th day and later the patient got warfarin. In two months after surgery INR was 2,2-2,5, antithrombin III – 75%, thrombocytes 373*109 /l, D-dimer – positive (>250 ng/ml), however, venous thrombosis were not disclosed.
Conclusion: Complex approach with use of modern medication allows improving treatment results of patients with congenital heart diseases with presence of homeostasis pathology.