Acute systemic-pulmonary shunt occlusion in cyanotic congenital heart diseases: Are hereditary thrombophilic factors really matter?

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Objective: Acute thrombosis of systemic to pulmonary shunts (SPS) in cyanotic congenital heart disease is a crucial complication for patients have shunt dependent pulmonary blood flow. Our aims were to determine the outcomes in patients early after undergoing SPS operations and to assess the presence of inherited thrombophilia factors in the children experienced occlusion of SPS.

Material and methods: A prospective randomized clinical trial was performed in a tertiary referral congenital cardiac center. Patients with hypoplastic left heart syndrome were excluded. The patients experienced shunt thrombosis were assessed for inherited and acquired thrombophilic factors. Vitamin B12 and folic acid levels and antithrombin III, protein C (PC), protein S (PS), anti-cardiolipin antibody (ACA), homocysteine, factor 5 Leiden and prothrombin G20210A mutations were studied.

Results: From October 2010 to September 2012 seventy-seven children underwent first shunt operation with a median age of 61 days (range: 4 days-5,6 years) and median weight of 8,8 kg (range: 2.1-14 kg). Thirty-three (%43) of them were neonate. The shunt sizes varied from 3 to 5 mm. The rate of acute shunt blockage was 10% (8/77), all within the first 24 h. There were 24 hospital deaths (31%), three of them was associated with shunt occlusion. There were 3 hereditary thrombophilia (1 antiphospholipid syndrome, 1 protein C deficiency, 1 active protein C resistance) in patients who had shunt thrombosis.

Conclusion: SPS occlusion can be potentially life-threatening in the case of patients have shunt dependent pulmonary blood flow. When shunt occlusion occurred especially in patients with recurrent shunt thrombosis, inherited thrombophilia factors should be investigated.