The hematocrit – an important factor causing impaired haemostasis in patients with cyanotic congenital heart disease

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Introduction:
Patients with cyanotic congenital heart disease (CCHD) have haemostatic abnormalities, which result in an increased risk of bleeding. The cause is unknown, but recent studies have indicated that an elevated hematocrit, which is present in cyanotic patients, could be an important factor. This is contrary to the previous assumption that elevated hematocrit is associated with increased risk of thrombosis. The aim of this study was to characterize haemostatic abnormalities, examine how changes in hematocrit affect the haemostatic profile and whether a hematocrit reduction could terminate bleeding in CCHD patients.

Methods: This was a prospective, multicenter study. The haemostatic profile was characterized in ninety-seven clinically stable CCHD patients. To evaluate the influence of hematocrit on the haemostatic profile, 21 of the patients underwent phlebotomy and 16 patients received treatment with an iron supplement. Furthermore ten patients with hemoptysis underwent phlebotomy. The haemostatic profile was reevaluated after interventions. The haemostatic profile consisted of hematocrit, platelet count, thrombelastograph (TEG), and plasma coagulation analysis.

Results:
TEG revealed that patients with CCHD and elevated hematocrit were hypocoaguable. Furthermore a positive correlation between elevated hematocrit and hypocoaguable was present. Interventions such as phlebotomy and treatment with supplemental iron causing significant hematocrit changes confirmed the correlation between hematocrit and the haemostatic profile. Finally a hematocrit reduction by phlebotomy successfully terminated the hemoptysis in all ten CCHD patients.

Conclusion:
Patients with CCHD and elevated hematocrit are hypocoaguable. Haemostasis seems to be affected by changes in hematocrit and phlebotomy may be a possible treatment of bleeding in CCHD.