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Impaired fibrinogen function in patients with cyanotic congenital heart disease

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Introduction:
Patients with cyanotic congenital heart disease(CCHD) have haemostatic abnormities associated with bleeding and thrombo-embolic events. These haemostatic abnormalities are not fully understood, but recent studies indicate that elevated hematocrit and fibrinogen function may be of importance.
The aim of this study was to characterize haemostatic abnormities in CCHD patients and examine how these are affected by increased hematocrit.
Methods:
In a prospective study fifty adult CCHD patients had hematocrit, platelet count, and fibrinogen concentration examined. Thrombelastography (standard TEG) values R (initiation of coagulation), Angle (clot build-up), MA (maximal clot strength), as well as TEG Functional Fibrinogen (TEG FF) assay evaluating fibrinogen function (FLEV) was performed. In addition TEG FF gave information about fibrinogen and platelet contribution to clot formation and strength.
Results:
Average hematocrit was 56±8% and platelet counts in the lower normal range. Standard TEG revealed prolonged R, reduced Angle but normal MA indicating a hypocoaguable condition with impaired clot formation but preserved clot strength. Interestingly the standard TEG R, Angle and MA values were correlated to elevated hematocrit, indicating that elevated hematocrit caused a hypocoaguable state. Despite of high levels of plasma fibrinogen in patients with elevated hematocrit, TEG FF demonstrated that FLEV was diminished particularly affecting clot formation but also strength. On the other hand the platelet function was normal despite of low platelet count.
Conclusion:
Patients with CCHD are hypocoaguable mainly due to impaired fibrinogen function. Despite a low platelet count, platelet function seems to be normal in CCHD patients. Haemostasis, and especially fibrinogen function, is negatively affected by elevated hematocrit.