

## Coagulation Abnormalities in Pediatric Patients with Congenital Heart Disease: A Literature Review

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### Abstract:

It has been recognized that patients with Cyanotic Congenital Heart Disease (CCHD) show significant bleeding tendency which can be secondary to coagulopathies in these patients. Some coagulation abnormalities are thrombocytopenia, factor deficiencies, fibrinolysis and Disseminated Intravascular Coagulation (DIC).

According to high prevalence of CCHD and major operations in these patients, the aim was to evaluate the coagulation abnormalities in children with CCHD.

**Key words:** Coagulation Abnormalities, CCHD, Congenital Heart Disease, Pediatrics.

### Introduction

Patients with congenital heart disease susceptible to develop coagulation abnormalities. Several coagulation abnormalities reported such as low platelet, factor deficiencies, fibrinolysis and DIC (1).

Very little information is available about coagulation abnormalities in patients with congenital heart disease, but some patient with congenital heart disease with cyanosis are at high risk for coagulation disorder (2).

Thrombocytopenia and suppressed platelet aggregation are known factor underlying the bleeding tendency in patients with cyanotic congenital heart disease and Eisenmenger syndrome (5). A variety of coagulation disorders recognized and responsible

causative factors have been postulated to contribute, including hyper viscosity, DIC, and primary fibrinolysis(1). Other causative factor are platelet function abnormalities, decrease production of coagulation factor due to impaired liver function and vitamin K deficiency (1).

### Discussion

Over the 50 years multiple defect coagulation and fibrinolytic systems have been reported in children with congenital heart disease (3).

In 2011 Haroled and co-worker, evaluated 37 cyanotic and 28 acyanotic children with CHD. Four children with rheumatic valvular disease and one child with hyperthyroid heart disease. Impaired aggregation was found in 14 of 37 patients (%37.8) with CCHD and in 4 of 28 (%14.3) acyanotic CHD (4).

In one study in Irland by Cazzaniag and co-worker, 109 patients with CHD, a cyanotic

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patients only demonstrate an impairment of the hepatic synthesis of coagulation factors, while cyanotic patients also have a chronic compensated DIC by lower Prothrombin time (PT), Antithrombin III (ATIII) activity and platelet (PLT) count (3).

In Horigome studies, coagulation laboratory tests were performed on 65 children with CHD. In 37.8% of children with cyanotic CHD and 14% with a cyanotic, platelet aggregation was impaired. Impairment in aggregation was clearly correlated with the severity of hypoxemia and polycythemia in the cyanotic group (5).

In one of study in Sweden, Henriksson evaluated 41 cyanotic patients and concluded that haemostatic abnormalities were common. The defects can be explained by deficient synthesis resulting from systemic hypoxia as well as from sluggishness of the local microcirculation caused by high blood viscosity (3).

In one study in 2011 Zonguldak Turkey, 49 children who had CHD and needed surgical intervention were evaluated about coagulation abnormalities, 16 patients had prolonged PT, 13 patients had low fibrinogen level, 10 patients had prolonged activated Partial Thromboplastin Time (aPTT) and 5 patients had low platelet count(6).

In Goel study in New Delhi evaluated haemostatic changes in 20 children with cyanotic and 12 acyanotic patients with congenital heart disease in 1998. It was concluded that laboratory abnormalities of tests of hemostasis were more common in cyanotic group. The patterns of laboratory abnormalities suggested a chronic compensated disseminated intravascular coagulation at a subclinical level (7).

In one of study in Egypt in 2012, by Ismail and co-workers, were evaluated 23 children with cyanotic and 30 patients with acyanotic congenital heart disease and 30 healthy control group about platelet-derived microparticles and platelet function

profile. Von Willebrand factor antigen (vWF Ag) as a marker of endothelial dysfunction. Hemoglobin, hematocrit (HCT), D-dimer, and vWF Ag were significantly higher in CCHD than adult congenital heart disease group(ACHDG). Platelet Microparticles and P-selectin expression were increased in patients than controls, particularly in CCHD and positively correlated to HCT, D-dimer, and vWF Ag while platelet count, aggregation, and GP IIb/IIIa expression were decreased in CCHD compared with ACHD group and negatively correlated to HCT (8).

In one study in Los Angeles in 2006, Lill were evaluated pathogenesis of thrombocytopenia(PT) in 105 patients with cyanotic congenital heart disease. 26 patients had platelet counts lower than 100000 and non of them had platelet dysfunction (9).

In study with Nieble in Milwaukee in 2012 about thromboelastography in the assessment of bleeding following surgery for 60 patients with congenital heart disease. Pediatric patients with significant postoperative bleeding after surgery were more likely to have abnormal thromboelastography early after cessation of cardiopulmonary bypass (10).

In one study of Osthaus in Germany in 2008 on Whole blood coagulation in 51 infants with congenital heart diseases. Pathological thromboelastography parameters were found in seven (41%) cyanotic patients and in three (17%) acyanotic patients and more than one abnormal thromboelastography coagulation parameter was found in four patients, all of them cyanotic patients (11).

In one of the studies in 1987 with Colon-Otero on preoperative evaluation of hemostasis in 235 patients with congenital heart disease, preoperatively, the prothrombin time, partial thromboplastin time, activated partial thromboplastin time,

thrombin time, or platelet count were abnormal in 45 of the 235 patients (19%), a significantly higher incidence than that expected in a normal population (12).

### Conclusion

Over production of platelet microparticles and suppression platelet aggregation, both related to hyper viscosity, may play an important role in the pathogenesis of hemostatic defect and coagulation abnormalities in patients with CCHD.

Abnormal haemostatic test results are common in patients with congenital heart disease and caution must be taken during operation of these patients.

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