HAEMOSTATIC DISORDERS IN CHILDREN WITH CONGENITAL CYANOTIC HEART DISEASES

Thesis

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ABSTRACT

Patients with cyanotic congenital heart disease (CCHD) and associated secondary polycythemia are susceptible to develop coagulation abnormalities. Our study included one hundred patients with CCHD, 37% had TOF, 35% had TGA and other CCHD as TAPVR, single ventricle and pulmonary atresia. The age was between 4 days and 16 years. There was a statistically significant positive (direct) correlation between age, Hb% and HCT. Also, there was a statistically significant positive (direct) correlation between O2 and platelet count. We found that the indications of partial exchange transfusion were polycythemia, symptoms or both and it improved the platelet count so we recommend PET for patients with CCHD who are associated with secondary polycythemia, thrombocytopenia or other coagulation defects as a preoperative preparation for cardiac surgery because it will decrease the risk of postoperative bleeding in those patients. The study showed that patients with congenital cyanotic heart diseases may need pre and post operative management as vitamin k, FFP, platelet transfusion to improve the coagulation function and decrease the risk of post operative bleeding.

Keywords:

CCHD- PET- hemostatic disorders- thrombocytopenia

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To my Mother and Father

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ABBREVIATIONS

ACT	The activated clotting time
APTT	Activated partial thromboplastin time
AT	Antithrombin
BSA	Body surface area
BT shunt	Blalock-Taussig shunt
CCHD	Cyanotic congenital heart disease
CHD	Congenital heart disease
CNS	Central Nervous System
CPB	Cardiopulmonary bypass
CT	Computerized Tomography
DIC	Disseminated Intravascular Coagulation
DVT	Deep Vein Thrombosis
EACA	ϵ-aminocaproic acid
ELT	The euglobulin ck lysis time
FDPs	Fibrin degradation products
FFP	Fresh frozen plasma
Hb	Hemoblobin
HCT	Hematocrite
ICU	Intensive care unit
MCH	Mean corpuscular hemoglobin
MCV	Mean corpascular volume
MRA	Magnetic resonance angiography
MRI	Magnetic resonance imaging
OR	Operation room
PA	Pulmonary angiography
PAI	Plasminogen activator inhibitor
PC	Prothrombin concentration
PE	Pulmonary embolus
PET	Partial exchange transfusion
PF	Platelet Phospholipid
PLT	Platelet
PT	Prothrombin time
PTT	Partial thromboplastin time
SD	Standard deviation
TA	Tranexamic acid
TAPVR	Total anomalous pulmonary venous return

TF	Tissue factor
TFPI	Tissue factor pathway inhibitor
TGA	Transposition of great arteries
TOF	Tetralogy of fallot
TT	Thrombin time
US	Ultrasonography
VSD	Ventricular septal defect

INTRODUCTION

The incidence of congenital heart disease (CHD) is approximately six to eight in 1,000 live births. While the majority of these infants are born with mild disorders, 15 percent are potentially life-threatening and a quarter of these infants are discharged after their birth hospitalizations without being diagnosed. Of newborns with potentially fatal forms of CHD, one-third have cyanotic lesions. Early recognition, emergent stabilization, and transport to an appropriate cardiac care center are critically important in the outcome of these patients **(Wren** *et al.,* **2008).**

Polycythemia in patients with cyanotic heart disease can cause a hyperviscosity state. These patients may have headache, increased cyanosis, decreased exercise tolerance, and stroke. The only treatment is removal of red cells by phlebotomy, partial-exchange transfusions, or erythrocytapheresis **(James** and **Corrigan, 2008).**

Patients with cyanotic congenital heart disease (CCHD) and associated secondary polycythemia are susceptible to develop coagulation abnormalities. Several coagulation defects, including thrombocytopenia, factor deficiencies, fibrinolysis, and disseminated intravascular coagulation (DIC), have been reported in these patients **(Depaak** and **Virmani, 2002).**

CCHD is associated with thrombocytopenia. The etiology is unclear, but the mechanism appears to involve decreased production of megakaryocytes **(Lill** *et al.,* **2006).**

There is an inverse relationship between the blood oxygen saturation and the hemoglobin in patients with CCHD, especially after the age of 3 years. Thus, the degrees of cyanosis and polycythemia dictate the presence of the thrombocytopenia. The thrombocytopenia is rarely severe. Additionally, the platelets do not function normally in vitro tests. Thus, the platelet defect in patients with CCHD is both quantitative and qualitative. Spontaneous bleeding is unusual, but bleeding may be excessive with injury and surgery. The treatment is to correct the polycythemia and hypoxia, which allows the platelet count and function to return to normal. Platelet transfusions are rarely needed. Corticosteroids and intravenous globulin therapies are not helpful **(James** and **Corrigan, 2008).**

Extracorporeal circulation introduces additional problems with regard to blood coagulation and may further interfere with an already faulty clotting mechanism, thereby aggravating the likelihood of hemorrhage in these patients. On the other hand it may be beneficial by providing an exchange transfusion of fresh blood which contains necessary clotting factors. As a result of these conflicting effects it is difficult to predict the occurrence of hemorrhage after the use of extracorporeal circulation **(Fletcher** *et al.,* **2000).**

Given that definitive surgical correction of congenital lesions in neonates involves complex repairs with a long duration of cardiopulmonary bypass (CPB) and multiple extracardiac suture lines, these coagulopathies predispose the patient to increased bleeding and transfusion requirements perioperatively **(Guay** and **Rivard, 1996).**

It is reasonable to believe that avoiding excessive hemorrhage after cardiac surgery with CPB in these patients is an important factor in improving surgical outcome. Understanding the management of hematologic disorders that accompany CCHD is an important clinical priority for the cardiac anesthesiologist. Therapeutic guidelines have focused primarily on the management of polycythemia*;* however, other controversies exist **(Depaak** and **Virmani, 2002).**

The present work presents the pathophysiology of coagulation abnormalities in these patients and current management strategies.

AIM OF THE WORK

The present work aims at*:*

- Identifying the prevalence, risk factors and lines of management of hemostatic disorders among children with CCHD.
- Study the effect of partial exchange transfusion on the platelet count and other coagulation functions in patients with CCHD.
- Study the risk factors, lines of management of postoperative bleeding and changes in coagulation function in the patients with CCHD who undergo cardiac surgeries.