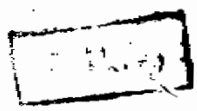
HEMOSTASIS IN CONGENITAL & RHEUMATIC HEART DISEASE



AN ESSAY

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BY

SAYED ABDALLA SAYED

M.B., B.Ch.

SUPERVISORS

Prof. Dr. SAWSAN EL-SOKKARI

Prof. of Pediatrics

Faculty of Medicine

Ain Shams University

Dr. HODA LOTFI EL-SAYED

Lec. of Pediatrics

Faculty of Medicine

Ain Shams University

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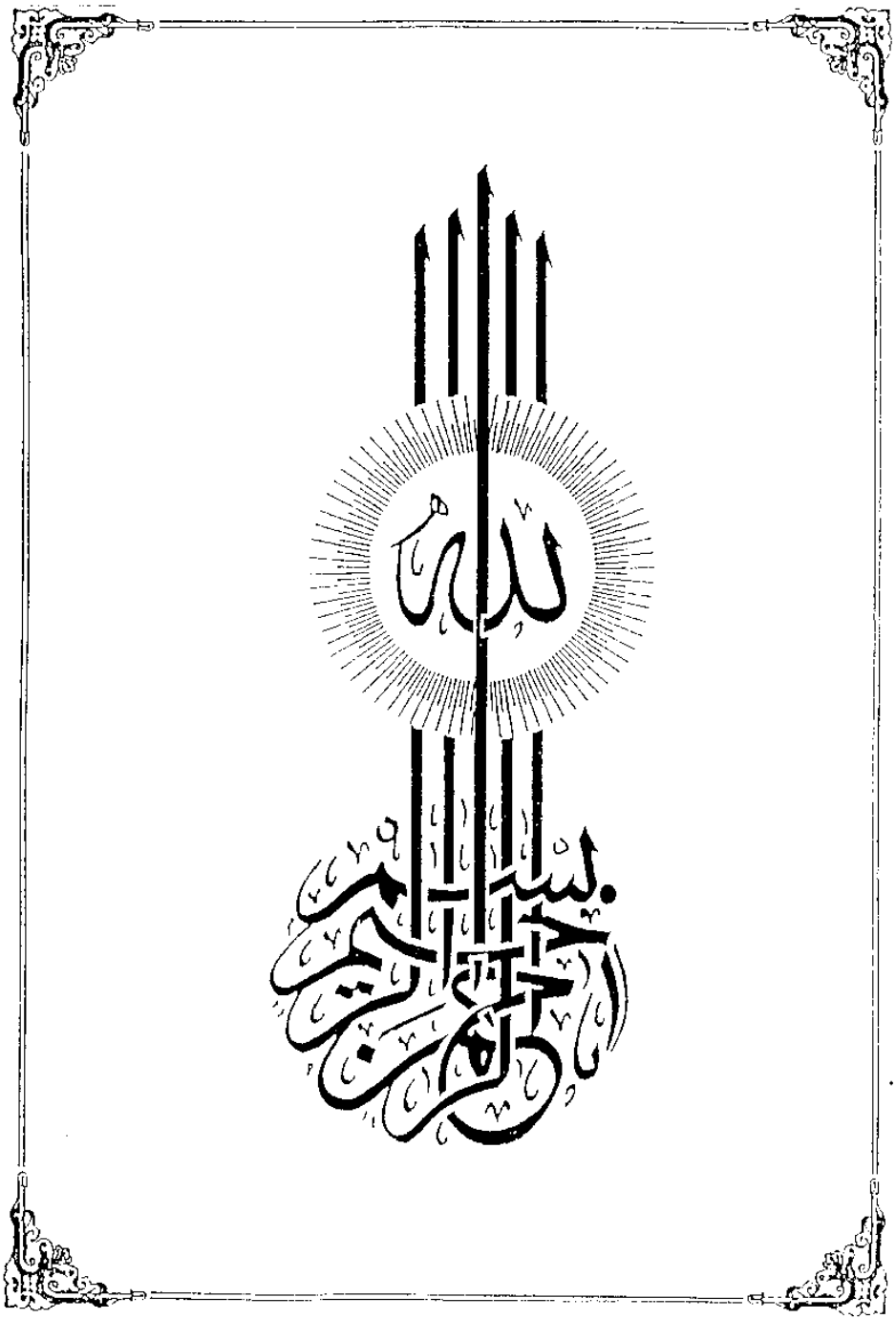

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LIST OF ABBREVIATIONS

C.H.D. : Congenital Heart Disease.

R.H.D. : Rheumatic Heart Disease.

M.S. : Mitral Stenosis.

A.S. : Aortic Stenosis.

A.S.D. : Atrial Septal Defect.

V.S.D. : Ventricular Septal Defect.

R.V.H. : Right Ventricular Hypertrophy.

L.V.H. : Left Ventricular Hypertrophy.

C.V.H. : Combined Ventricular Hypertrophy.

V.W.F. : Von Willibrand factor.

m-R.N.A. : Messenger Ribonucleic Acid.

U-PA. : Urokinase Plasminogen Activator

t- PA : Tissue Plasminogen Activator.

H.M.W.K. : High Molecular Weight Kininogen.

PF-3 : Platelet Factor - 3

PDGF : Platelet Derived Growth Factor.

A.D.P. : Adenosine Diphospahte.

A.T.P. : Adenosine Triphospahte.

A.A.T. : Alpha-1- Antitrypsin.

A.T.III : Antithrombin III.

PC : Protein C

APC : Activated Protein C.

PGs : Prostaglandins.

PGI_2 : Prostacyclins.

TXA_2 : Thromboxane A_2

D.I.C. : Dissiminated Intravascular Coagulopathy.

D.D.A.V.P.: 1-Desamino, 8-D-Arginine Vasopressin.

B.T. : Bleeding Time.

T.T. : Thrombin Time.

S.T.T. : Serial Thrombin Time.

P.T. : Prothrombin Time.

P.T.T. : Partial Thromboplastin Time.

A.P.T.T. : Actiated Partial Thromboplastin Time.

E.L.T. : Euglobulin Lysis Time.

F.D.P. : Fibrin Degradation Products.

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INTRODUCTION

Hemostasis is the cessation of bleeding following trauma to blood vessels, it results from 3 processes: first, contraction of vessel walls, secondly, formation of a platelet plug at the site of the break in the vessel wall, and thirdly, the formation of a fibrinclot.

The commonest cause of bleeding is that resulting from a deficiency of platelets and the second commonest cause is an abnormality in the clotting mechanism, the last cause is vascular abnormalities.

Recently, new factors in the clotting mechanisms, comes to light such as Prostaglandins & Protein C. [Luscher,1987].

Bleeding may not occur until the balance is disturbed further by trauma or some other events as fever, infections or anemia. The situation is more complex when hemostasis is disturbed as a result of systemic illness with associated metabolic upsets, hypoxia, disordered circulation and variable degree of organ failure [Castaldi, 1987].

Congenital heart diseases occur in approximately 8/1000 live births. Approximately one third of all cases of C.H.D. are cyanotic. Two common consequences of the cyanosis are polycythemia and finger clubbings. Marked polycythemia is associated with high hematocrit value which leads to increased viscosity and adds to the work of the heart and increase risk of arterial and venous thrombosis [Nora, 1971].

Acute rheumatic fever and rheumatic heart diseases remain serious medical problems , while the incidence of acute rheumatic fever and rheumatic heart disease started to decline in developed countries, its incidence is still high in developing and 3rd world countries. [Schulman, 1984].

The clotting characteristics in rhuematic heart diseases are variable, in patients with mitral valve diseases, aggregation of platelets is significantly greater in pulmonary than in systemic arterial blood at rest, the converse is true during exercise. In aortic valve diseases, platelets aggregation is greater in systemic than in pulmonary arteiral blood at all times.

In patients with rheuamtic mitral valve disease and aortic valve disease, there is an increased thrombotic tendency in blood in the left heart which is particularly pronounced during exercise [Toy et al.,1980].

AIM OF THE WORK

The aim of this essay is to review the different references discussing the normal hemostasis, different congenital and Rheumatic heart diseases in infants and children and the associated hemostatic defects and factors affecting hemostasis in such patients.

PHYSIOLOGY OF HEMOSTASIS

PHYSIOLOGY OF HEMOSTASIS

Hemostasis is the prevention of blood loss. Capillaries and arterioles are ruptured continuously by the minor traumas of everyday life, and hemostatic mechanisms keep blood loss to a minimum. These mechanisms are crucial for survival when blood loss is appreciable. Their importance is especially obvious in patients who have defective hemostatic systems, anything more than minor vascular trauma may cause severe life threatening hemorrhage in the patients. [Colman, et al.,1982].

Events in Hemostasis:

Hemostasis is divided into the following phases:

1. Vascular phase.
2. Platelet phase.
3. Plasma phase.

1. Vascular phase

Vasoconstriction is the immediate response to vascular injury. The factors involved are contraction of vascular smooth muscle in direct response to injury, vasoconstriction in response to pain, and some vascular compression by the pressure of the blood lost into the surrounding tissues. The value of these immediate responses is especially apparent in

cases of severely injuried small blood vessels. The ability of vessels as large as the radial artery to constrict immediately can decrease blood loss significantly [Wintrobe et al.,1981].

In general, this mechanism is most effective in vessels damaged by blunt instruments (chains, bricks, or gun shot wounds) and is less effective when wounds are made by sharp objects (knives, broken bottles , or ice picks). [Lee and Boggs, 1981].

Vessels with muscular coat contract following injury, thus assisting hemostasis plug formation by reducing blood flow. Vaso-constriction occurs, however, even in the microcirculation in vessels without smooth muscle cells due to the release of vasoactive substances from the platelets which are serotonin and thromboxane A_2 [Hamberg et al., 1975].

The endothelial cells play an active role in hemostasis as they contract following injury or exposure to bradykinin, serotonin and histamine. It also synthesis and secretes at least three substances which are involved in the formation and localization of the hemostatic plug. These are Von Willebrand factor, Prostacyclin and Plasminogen activator [Stemmerman,1974].

Von Willebrand factor (VIII: VWF) is a part of a moleccomplex which also possesses factor VIII clotting activity [Bloom,1977].