IMMUNOLOGICAL ASPECT IN COAGULATION DISORDERS
(ESSAY)

THESIS

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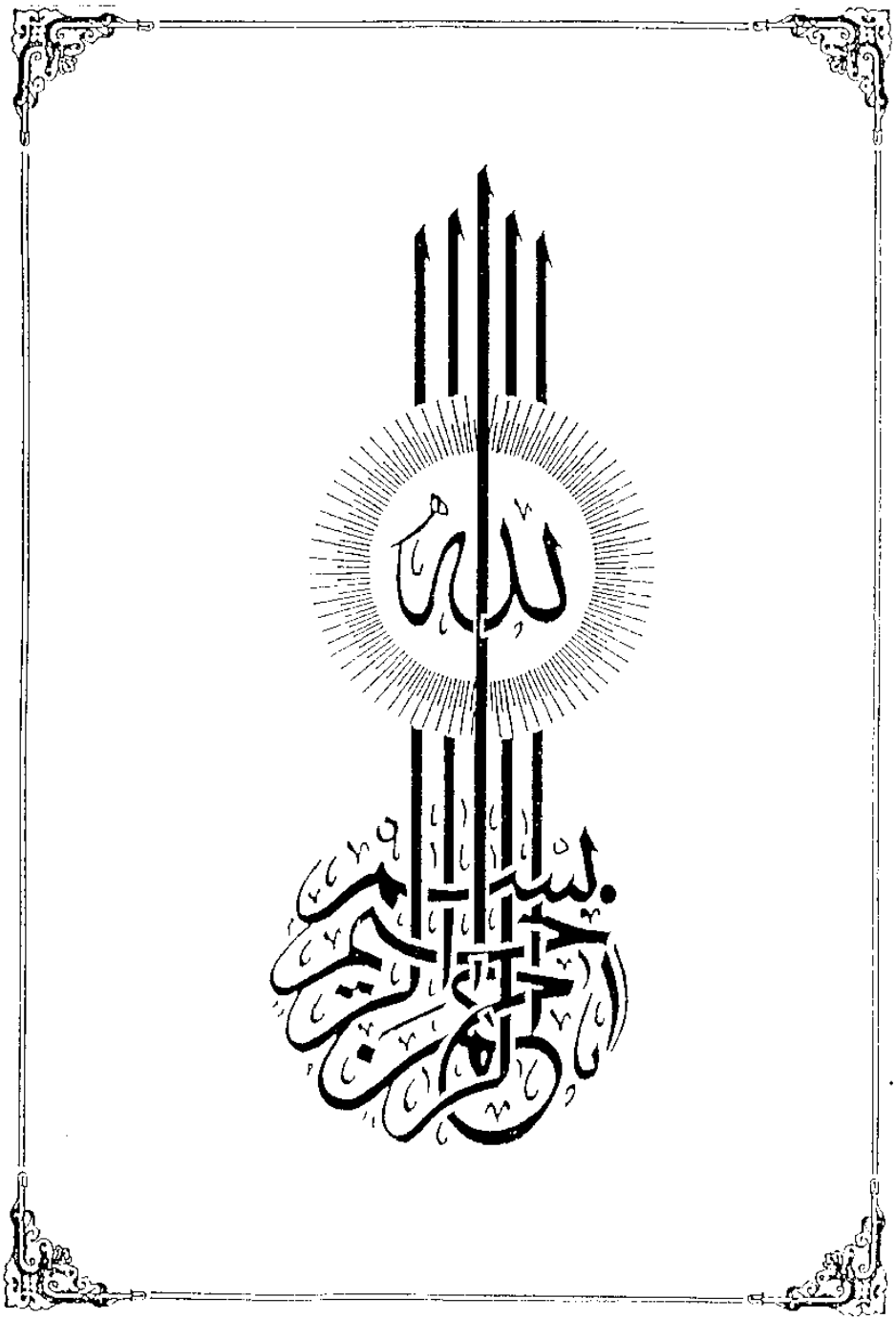
بسم الله الرحمن الرحيم

" قالوا سبحانك لا علم لنا الا ما علمتنا
انك أنت العليم الحكيم "

صدق الله العظيم

سورة البقرة : آية ٣٢



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- F.D. P = Fibrin degradation products.
- PTT = Partial thromboplastin time.
- VIII:C = Antihemophilic factor procoagulant activity
- TGT = Thromboplasten generation test.
- AIDS = Acquired immune deficiency syndrom
- FFP = Fresh frozen plasma.
- Cryo. = cryoprecipitate
- DDAUP = 1-deamino 8-d- arginine vasopressin
- FEIBA = Factor VIII inhibitor by passing activity
- APCCS = Activated prothrombin- complex concentrates
- DIC = Disseminated intravascular coagulation.
- PA. IgM= platelet associated IgM.
- PA.IgG = platelet associated IgG.
- ITP = Idiopathic thrombocytopenic purpura.
- VIIIR: Ag= Factor VIII related antigen
- HMW kininogen = High molocular weight kininogen.

CONTENTS

INTRODUCTION

Introduction

Coagulation disorders are classified into three groups: Coagulation disorders due to deficiency of the coagulation factors, due to platelet destruction or due the vascular abnormalitis. The immunological aspect in these disorders are helpful in the managment of such cases. In cases of hemophilia the detection of factor VIII inhibitors are helpful in managed such cases. The immunological study is helpful in diagnosis, management and prevention of different disease. In ITP detection of IgG and IgA are essential to know the type of ITP and for proper managment of such cases.

AIM OF THE WORK

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The aim of this review is to classify and discuss the different types of coagulation disorders. The study of the immunological aspect of the coagulation disorders are helpful in proper diagnosis as in case of ITP and proper managment e.g. proper management of hemophilic patients with inhibitors specific to factor VIII.

BLOOD COAGULATION

Blood coagulation

The hemostatic process has three major components:

1- Integrity of the vascular wall and reaction of the blood vessels to injury.

2- Platelet activity, and

3- Coagulation of the blood (Philip, . . 1980).

Nomenclature of blood coagulation factors :

The problems of nomenclature have been partially solved by the development of an international standard nomenclature (Macfarlane, 1976).
Table (1) gives this recommended nomenclature.

Coagulation mechanism devided into three phases : phase I deals with the formation of prothrombin activators and can be further subdivided into the intrinsic and extrinsic clotting systems. Phase 2involves the cleavage of prothrombin into intermediates, one of which is thrombin. During phase 3 fibrinogen is converted to fibrin, and the clot is stabilized.

Table (1) Nomenclature and Synonyms for Coagulation Factors

Roman Numeral	*Preferred Descriptive Name*	*Synonyms*
I	Fibrinogen	
II	Prothrombin	
III	Tissue factor	Thromboplastin
IV	Calcium ions	
V	Proaccelerin	Labile factor, accelerator globulin (AcG), thrombogen
VII	Proconvertin	Stable factor, serum prothrombin conversion accelerator (SPCA)
VIII	Antihemophilic factor (AHF)	Antihemophilic globulin (AHG) antihemophilic factor A, platelet cofactor 1, thromboplastinogen
IX	Plasma thromboplastin component (PTC)	Christmas factor antihemophilic factor B, autoprothrombin II, platelet cofactor 2
X	Stuart factor	Prower factor, autoprothrombin III, thrombokinase
XI	Plasma thromboplastin antecedent (PTA)	Antihemophilic factor C
XII	Hageman factor	Glass factor, contact factor
XIII	Fibrin stabilizing factor (FSF)	Laki-Lorand factor (LLF), fibrinase, plasma transglutaminase, fibrinoligase
—	Prekallikrein	Fletcher factor
—	HMW Kininogen	High molecular weight kininogen, contact activation cofactor, Fitzgerald factor, Williams factor, Fleujeac factor, Reid factor, Washington factor

(Wintrobe at al., 1981)

Phase I: Intrinsic clotting system

The intrinsic system is responsible for the formation of a clot in plasma from which all tissue juices are excluded and from which blood cells other than platelets are removed. It involves factors XII, XI, X, IX, VIII, and V, platelet factor 3 and calcium. Clotting via the intrinsic system begins when factor XII is activated by contact with a foreign surface (Wilner et al., 1968). Factor XII a functions to activate factor XI. Neither activation of XII nor activation of XI requires calcium (james, 1975).

The next step in the intrinsic system is the activation of factor IX by the enzyme XIa, this reaction requires calcium, and IXa is a potent coagulant. The next reaction is a complicated one involving factor IX a, factor VIII, calcium, and the phospholipid derived from platelets, platelet factor 3 (Davie et al., 1969). The IX a - VIII - phospholipid complex activates factor X which in turn participates in the conversion of prothrombin to thrombin (Borton et al., 1967). Factor X a is the enzyme that is responsible for the cleavage of prothrombin and factor V and phospholipid only accelerate this reaction (jobin and Esnouf, 1967).