HELLP SYNDROME

THESIS

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BY

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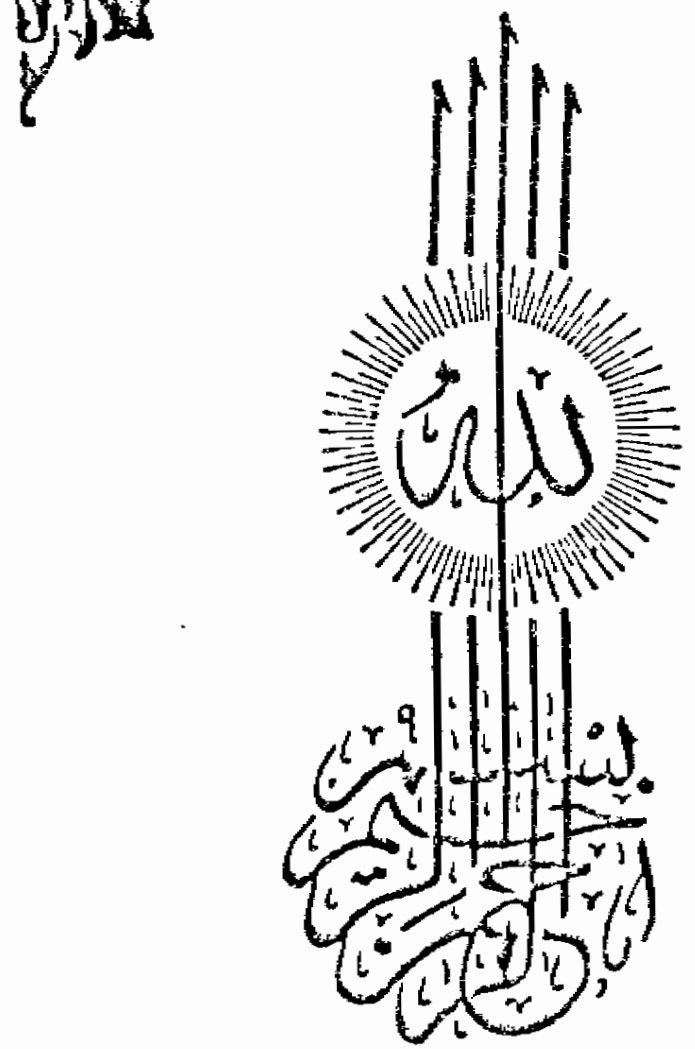

قَالُوا سُبْحَانَكَ لَا عِلْمَ لَنَا إِلَّا مَا
عَلَّمْتَنَا إِنَّكَ أَنْتَ الْعَلِيمُ الْحَكِيمُ .

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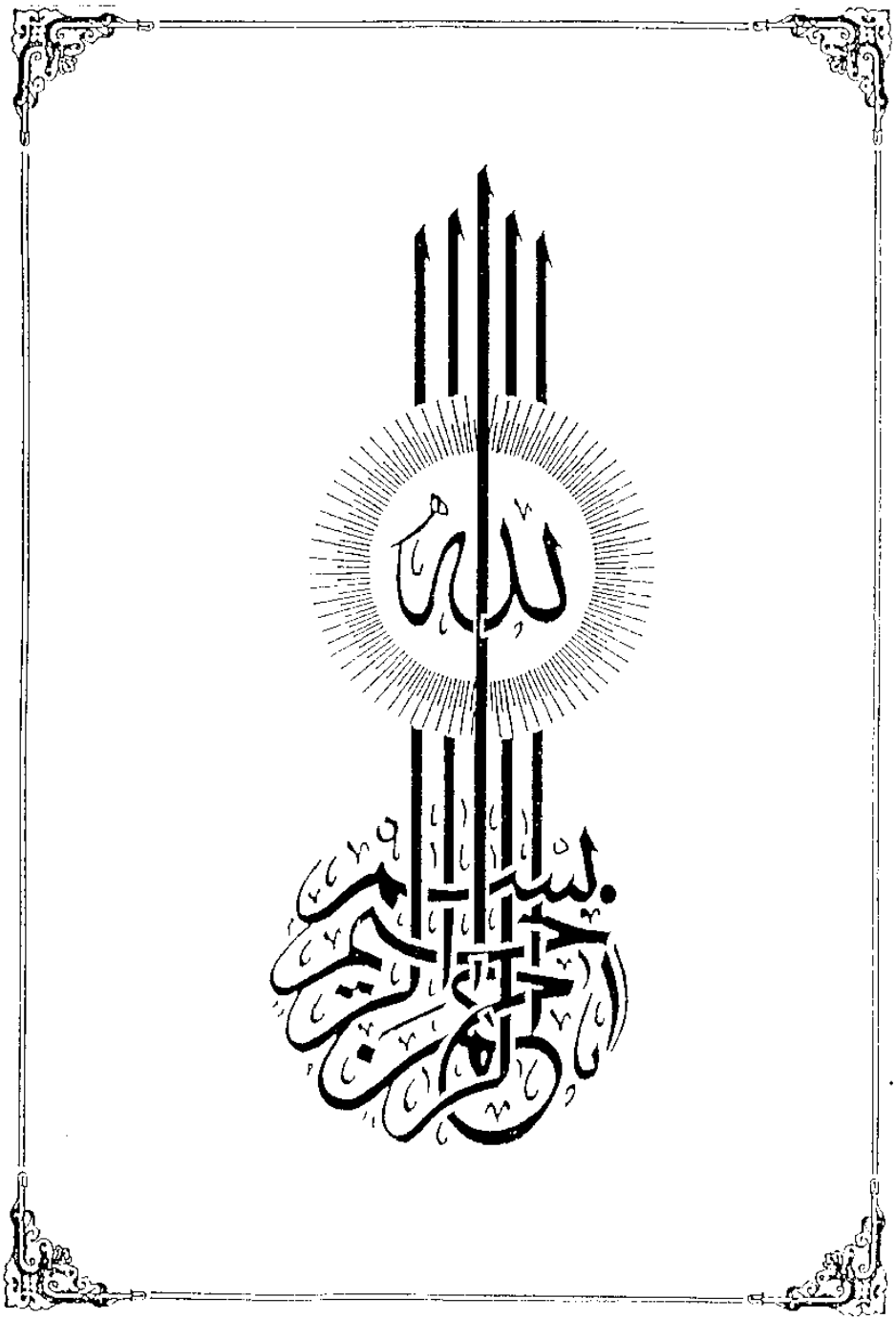

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CONTENTS

INTRODUCTION

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The hypertensive disorders of pregnancy represent one of the greatest challenges to the medical skills of the obstetricians. The syndrome of pre-eclampsia-eclampsia complicates about 7% of all pregnancies and is a major cause of maternal and perinatal morbidity and mortality worldwide . Hogberg (1986) .

Sibai in 1989 reported that hypertension is diagnosed in a wide spectrum of patients, ranging from those who have only a minimal elevation in blood pressure to those who experience severe hypertension with multiple organ dysfunction . The manifestation of hypertension in these patients are clinically similar but result from several different pathologic processes with various underlying causes. The two most common forms of hypertension are pregnancy-induced hypertension (P.I.H.), a disorder that appears during pregnancy and is reversed by delivery, and preexisting chronic hypertension. The latter condition is unrelated to but coincides with pregnancy, may be detected for the first time in pregnaney, and is not reversed by delivery. In certain cases the two conditions may occur together in one patient [superimposed P.I.H. or preeclampsia] . The entity of P.I.H. is responsible for approximately 70% of all hypertension, while chronic hypertension accounts for the remaining cases .

The ultimate challenge to obstetricians is to be able to detect the disease in its early stage, especially in those cases developing in mid-trimester and in those with an atypical presentation. The question of whether the HELLP syndrome exists as a distinct entity or is part of a spectrum of pregnancy complication, which have in common hemolysis, elevated liver enzymes and thrombocy-topenia has long been a source of speculation and debate among obstetricians and internist. How ever Sibai (1990) through his review of the literature found that there is a definite need for a uniform definition, diagnosis and management of this syndrome .

Hemolysis, abnormal liver function tests, and thrombocy topenia have been recognized as complications of preeclampsia-eclampsia for many years. Killam, et al (1975).

Weinstein and his colleuge in 1982 described 29 cases of severe preeclampsia-eclampsia complicated by thrombocyto penia,abnormal peripheral smear, and abnormal liver function test results. He suggested that this collection of signs and symptoms constituted an entity separate from severe preeclampsia and coined the term HELLP syndrome [H for hemolysis, El for elevated liver. enzymes and LP for low platelets]

Since then, several articles and case reports claiming to describe this syndrome have appeared in the medical lite-

rature. In addition, the presence of this syndrome became a major cause of litigation against obstetricians involving cases of alleged misdiagnosed preeclampsia. Controversy surrounds almost every aspect of the HELLP syndromp. Areview of the literature highlights the confusion concerning the terminology, incidence, cause, diagnosis and management of this syndrome. Sibai et al (1986) .

AIM OF THE WORK

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1 - Studying of the prevalance of HELLP syndrome in pregnancy induced hypertension .

2 - Correlation of HELLP syndrome with severity of the condition and fetal outcome .

TERMINOLOGY AND DIAGNOSIS

TERMINOLOGY AND DIAGNOSIS :

The terminology and diagnostic criteria used to describe this syndrome have been confusing and inconsistent. Goodlin in (1982) considered it an early form of severe preeclampsia and labeled it as a great imitator, impending gestosis. EPH [edema, proteinuria, hypertension] gestosis type B, and expanded toxemia syndrome.Weinstein (1982) considered it a unique variant of precclampsia while Mackenna et al (1983) considered it as misdiagnosed preeclampsia .

Sibai (1990) through his studies about HELLP syndrome found that there are considerable differences regarding the time of onset, type and degree of laboratory abnormalities used to make the diagnosis of this syndrome. Patients may be represented with the abnormalities on admission, others may develop the abnormalities during the conservative mangement of preeclampsia, and others may develop the abnormalities in the postpartum period. Patients may represent with no evidence of hemolysis, hence these patients will fit the criteria for ELLP syndrome. Even in cases where hemolysis is present, the diagnosis is based mostly on the presence of an abnormal peripheral smear (burr cells or schistocytes). As regard the platelets count sibai (1990) found that diagnosis of thrombocytopenia may be based on count $< 150 \times 10^3 / mm^3$, $< 100 \times 10^3 / mm^3$ or $< 75 \times 10^3 / mm^3$. There is no consensus in the literature regardeng which liver function test abnormal-

ities should be used to diagnose the syndrome . However . Weinstein in 1982 reported abnormal serum glutamic oxaloacetic transaminase (S.G.O.T.), abnormul serum glutamic pyruvic transaminase (S.G.P.T.), and abnormal bilirubin values, however, levels were not stated. In addition, he made no mention of lactic dehgdrogenase (L.D.H.) as a diagnostic feature .

But Mackenna and associates in (1983) stated that liver enzymes were elevated in all their patients but didnot specify the level or identity of the enzymes measured .

Goodlin and Thiagarajah and associates in 1982 considered S.G.O.T. to be abnormal at al level of > 30 U/L. Brazy and associates in 1982 used a level of > 50 U/L and Sibai and associates in 1986 used a level of > 72 U/L . Vandam and associates in 1989 used a level of > 16 U/L. In addition, the threshold for L.D.H. levels was variable among these studies (195 to 600 U/L).

However, recent reports suggested that all patients with HELLP syndrome will have D.I.C. if sensitive laboratory tests are used . Sensitive determinants of this condition include antithrombin III , fibrinopeptide A, fibrin monomer, D-dimer, plasminogen, and fibronectin. Unfortunately, these tests are expensive, time consuming, and not suitable for routine clinical monitoring . Vandam et al (1989) .

In consideration of the above problems, Sibai and associates in 1990 suggest that uniform and standardized laboratory values must be used to diagnose this Syndrome , also L.D.H. and bilirubin values should be included in the diagnosis of hemolysis. In addition, the degree of abnormality of liver enzymes should be defined as a certain number of standard deviation from the normal values for each hospital population. Furthermore , the rate of change in either liver enzymes or platelet count may be as important as the absolute value in establishing the diagnosis.

The criteria for the diagnosis of this syndrome require the presence of these laboratory findings :

(1) Hemolysis, defined by abnormal peripheral smear, increased bilirubin (> 1.2 mg / dI) and increased lactic dehydrogenase (> 600 U/L).

(2) elevated liver enzymes, defined as increased S.G. O.T. (> 70 U/L) and increased L.D.H.

(3) Low platelets, defined as platelet count < 100 x 10^3 / mm ^{3} Sibai (1990) .

In 1987 poldre concluded that in suspected cases of the HELLP syndrame in which evidence of hemolytic anemia is absent, assay of the serum haptoglobin may assist in the diagnosis.Haptoglobin is an α_2 plasma protein that binds to