Pregnancy Outcome in Patients with Budd Chiari Syndrome: A Single Centre Experience

Thesis

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بسم الله الرحمن الرحيم
وَأَنْزَلَ اللهُ عَلَيْكَ
الْكِتَابَ وَالْحِكْمَةَ
وَعَلَّمَكَ مَا لَمْ
تَكُنْ تَعْلَمُ وَكَانَ
فَضْلُ اللهِ عَلَيْكَ
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List of Abbreviations

Abb.	Full term
A	*Aorta*
AFP	*Alpha feto protein*
ALT	*Alanine aminotransferase*
ANA	*Antinuclear antibody*
APL	*Antiphospholpid*
AST	*Aspartate aminotransferase*
AZ	*Azygos vein*
BCS	*Budd chiari syndrome*
BCSG	*Budd chiari study group*
CALR	*Calreticulin gene*
CI	*Confidence interval*
CS	*Caesarean section*
CT	*Computed tomography*
DM	*Dibetes mellitus*
DNA	*Deoxyribonucleic acid*
DVT	*Deep venous thrombosis*
ET	*Essential thrombocytosis*
FDA	*Food and drug administration*
FVLM	*Factor v leiden mutation*
GA	*Gestional age*
GI	*Gastrointestinal*
HELLP	*H : hemolysis – el :elevated liver enzymes – lp: low platlats*
Hb	*Hemoglobin*
HBV	*Hepatitis b virus*
HCC	*Hepatocellular carcinoma*
HCV	*Hepatitis c virus*
HVOTO	*Hepatic venous outflow tract obstruction*
ICU	*Intensive care unit*
INR	*International normalization ratio*
IRHV	*Inferior right hepatic vein*

List of Abbreviations cont...

Abb.	Full term
IUD	*Intrauterine device*
IVC	*Inferior vena cava*
IVF	*In vitro fertilization*
LFT	*Liver function tests*
LIPV	*Left inferior phrenic vein*
LL	*Lower limb*
LMWH	*Low molecular weight heparin*
LRV	*Left renal vein*
MELD	*Model for End-Stage Liver Disease*
MPL	*Myeloproliferative leukemia virus oncogene*
MPN	*Myeloproliferative neoplasms*
MRI	*Magnetic resonance*
MTHFRD	*Methyl tetrehydro folate reductase*
NA	*Not applicable*
NICU	*Neonatal intensive care unit*
OCP	*Oral contraceptive pills*
PAP	*Pulmonary artery pressure*
PD	*Protein defiency*
PH	*Pulmonary hypertension*
PHG	*Portal hypertensive gastropathy*
PLT	*Platlats*
PT	*Prothrombin*
PTT	*Partial thromboplastin time*
PVT	*Portal vein thrombosis*
RUQ	*Right upper quadrant*
SAAG	*Serum ascites albumin gradient*
SD	*Standard deviation*
SLE	*Systemic lupus erythromatosis*
Smv	*Superior mesenteric vein*
SPSS	*Statistical package for social sciences*
SV	*Splenic vein*

List of Abbreviations cont...

Abb.	Full term
TIPSS	*Transjugular intrahepatic portosystemic shunt*
UFH	*Unfractionated heparin*
US	*Ultrasound*
US	*United states*
UTI	*Urinary tract infection*
WBC	*White blood cells*

INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare disorder caused by hepatic venous outflow obstruction and resulting hepatic dysfunction due to sinusoidal congestion, ischaemic injury to the liver and portal hypertension ***(Valla, 2003).*** The main mechanism for BCS is thrombosis of the hepatic veins or of the terminal portion of the inferior vena cava ***(Janssen et al., 2003).*** The diagnosis of BCS is based on the demonstration of a hepatic venous outflow tract obstruction. This obstruction can be accurately documented by non-invasive imaging such Doppler ultrasonography, computed tomography (CT) or magnetic resonance imaging (MRI). Doppler ultrasonography is regarded as the initial technique of choice and offers a high sensitivity and specificity ***(Bolondi et al., 1991).***

The exact prevalence of BCS is unknown but has been estimated as 1 per 100000 of the general population worldwide ***(Valla, 2003),*** with a higher prevalence being evident in developing countries such as India, Nepal, South Africa ***(Wang and Jones, 1996).*** BCS affects all races, usually during the third or fourth decades of life, and is more common in females ***(Valla, 2002).***

In Egypt the epidemiology of BCS showed that the presentation was chronic in 79.8% of patients, acute or subacute in 19.1% and fulminant in 1.1%. Factor V Leiden mutation (FVLM) was the most etiological cause of the disease

(53.1%) followed by mutation of the gene encoding for methyl tetra hydro folate reductase (MTHFR) (51.6%). Current or recent hormonal treatment was documented in 15.5% of females, aetiology couldn't be determined in 8.5% of patients. Males had significantly higher rates of MTHFR gene mutation and Bechet's disease, while females had significantly higher rates of secondary antiphospholipid antibody syndrome ***(Sakr et al., 2011).***

The management using stepwise regimens is largely successful with anticoagulation and interventional radiology alone. Stepwise regimen includes; anticoagulant therapy for an indefinite period; angioplasty or stenting for stenosis of hepatic veins; and decompressive techniques [surgical shunt or trans jugular intrahepatic portosystemic shunts (TIPSS)] for patients who are nonresponsive to medical treatment or not candidates for angioplasty/Stenting ***(EASL, 2016).*** TIPSS has a lower morbidity and mortality rate than surgery and is a preferred approach. The outcomes are favourable with 10-year survival approaching 90% ***(Tripathi et al., 2017).***

Patients with BCS usually have risk factors for venous thromboembolism. Pregnancy is a hypercoagulable state and earlier studies reported that women with BCS could be at risk of developing severe exacerbation of their underlying disease during pregnancy ***(Dilawari et al., 1994).*** Several previously reported observations suggest that pregnancy in BCS women in the West could cause deterioration of the liver disease and

pregnancy was associated with development of ascites in several women with known BCS ***(Martinelli et al., 2006).***

The desire for pregnancy is increasingly expressed by those young patients once their condition has greatly improved**.**

Data concerning pregnancy in women with BCS is scarce and thus there are no clear-cut guidelines for the outcome and management of pregnancy in patients with BCS ***(Valla, 2008).***

Aim of the Work

To assess the maternal and foetal outcome in a group of women who became pregnant while having BCS.

Chapter 1

BUDD–CHIARI SYNDROME (BCS)

Budd–Chiari syndrome (BCS) has been defined as an obstruction of the hepatic venous outflow tract without any cardiac disorder. The obstacle causing BCS may be located in the small or large hepatic veins or on the suprahepatic portion of inferior vena cava (IVC), but does not include sinusoidal obstruction syndrome/ hepatic veno-occlusive disease ***(Valla, 2009).***

Epidemiology:

There is no evidence for a difference in incidence of BCS between the West and East. Throughout the world, nearly all cases /HVOTO appear to be caused by hepatic venous obstruction,associated or not associated with involvement of parts of the IVC. India, Pakistan, the Middle East, the Mediterranean, and Europe share many characteristics including of the level of obstruction and individual causal factors. However, in China, involvement of the IVC may be common. Availability of the experts in noninvasive imaging techniques are essential for diagnosis of cases in different populations and to obtain reliable epidemiological data ***(Valla, 2009).***

Table (1) shows that prevalence of BCS in Europe has been relatively consistent [0.35–0.8 cases per million (pmi) per