

**تقييم نتاج توسيع الوريد الكبدي بالبالونة وتركيب
الدعاة الكبدية في المرضى المصريين المصابين
بمتلازمة بد - كيارى**

رسالة

توطئة للحصول على درجة الدكتوراه في طب المناطق الحارة

مقدمة من الطيب

أحمد سمير عبد المعطي

بكالوريوس الطب والجراحة

ماجستير طب المناطق الحارة

تحت إشراف

الأستاذ الدكتور / عفت عبد المنعم الفخاخ

أستاذ طب المناطق الحارة - كلية الطب- جامعة عين شمس

الأستاذ الدكتور / محمد أمين صقر

أستاذ طب المناطق الحارة - كلية الطب- جامعة عين شمس

الدكتور / مصطفى حامد عبد العليم

أستاذ مساعد طب المناطق الحارة - كلية الطب- جامعة عين شمس

الدكتور / سارة محمود عبد الحكم

مدرس طب المناطق الحارة - كلية الطب- جامعة عين شمس

الدكتور / محمد الغريب أبو المعاطي

أستاذ مساعد الأشعة التشخيصية - كلية الطب- جامعة عين شمس

كلية الطب- جامعة عين شمس

2013

OUTCOME OF PERCUTANEOUS TRANSHEPATIC BALLOON
ANGIOPLASTY (PTBA) WITH STENTING IN EGYPTIAN PATIENTS
WITH BUDD-CHIARI SYNDROME

Thesis

Submitted for Partial Fulfillment of the
Degree of M.D. in Tropical Medicine

By

Ahmad Samir Abdelmoaty
M.B.B.Ch., M.Sc. (Tropical Medicine, Ain Shams
University)

Under Supervision of

Prof. / Effat Abd El-Monem El-Fekhfakh

Professor of Tropical Medicine
Faculty of Medicine-Ain Shams University

Prof./ Mohammad Amin Sakr

Professor of Tropical Medicine
Faculty of Medicine-Ain Shams University

Dr./ Mostafa Hamed Abdel Aleem

Assistant Professor of Tropical Medicine
Faculty of Medicine-Ain Shams University

Dr./ Sara Mahmoud Abd El-Hakam

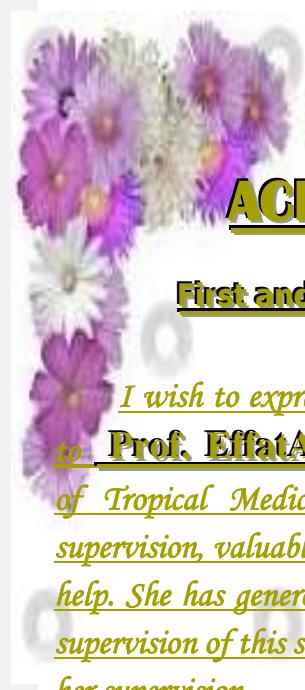
Lecturer of Tropical Medicine
Faculty of Medicine-Ain Shams University

Dr./ Mohammad El-Ghareeb Abou Elmaaty

Assistant Professor of Radiodiagnosis
Faculty of Medicine-Ain Shams University

Faculty of Medicine - Ain Shams University

2013



ACKNOWLEDGMENT

**First and foremost thanks to ALLAH,
the most merciful**

I wish to express my deep appreciation and sincere gratitude to Prof. Effat Abd El-Monem El-Fekhfakh, Professor of Tropical Medicine, Ain Shams University, for her close supervision, valuable instructions, encouragement and continuous help. She has generously devoted much of her time and effort for supervision of this study. It was a great honor to me to work under her supervision.

My deepest gratitude to Prof. Mohammad Amin Sakr, Professor of Tropical Medicine, Ain Shams University, for his close supervision, valuable instructions and continuous support. He gave me much of his valuable time, experience for planning and supervision of this study that cannot be expressed in words.

I wish to express my deep appreciation and sincere gratitude to Dr. Mostafa Hamed Abdel Aleem, Assistant Professor of Tropical Medicine, Ain Shams University, for his close supervision, valuable instructions and continuous help.

No words can fulfill the feeling of thanks I carry to Dr. Sara Mahmoud Abdelhakam, Lecturer of Tropical Medicine, Ain Shams University, for her continuous support and supervision.

My deepest gratitude to Dr. Mohammad El-Ghareeb Abou Elmaaty, Assistant Professor of

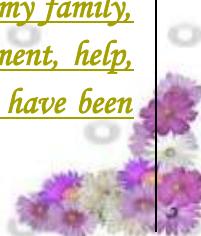
Radiodiagnosis, Ain Shams University, for his sincere guidance and valuable suggestions in the practical radiological part of this work.

Also, I wish to introduce my deep respect and thanks to Dr. Ahmed Kamal Eldorry, Professor of Radiodiagnosis and Dr. Mohammad Shaker Ghazy, Assistant Professor of Radio-diagnosis Ain Shams University for their great effort in this work.

Also, I must express my deepest thanks to all members of Budd Chiari Study Group (BCSG) and Egyptian Association for Study of Vascular Liver Disorders (EASVLD), who helped me in this work and patients who allowed me to produce this study.

Last, but not least, I would like to express my great thanks to Tropical Medicine Department and Interventional Radiology Unit, members and workers for their generously, help, and cooperation.

Finally I am particularly and deeply thankful to my family, for their continuous prayers, without their encouragement, help, understanding and moral support, this work could not have been achieved.



LIST OF CONTENTS

Title	Page No.
<i>Protocol of thesis</i>	
<i>List of Tables</i>	<i>I</i>
<i>List of Figures</i>	<i>VI</i>
<i>List of Abbreviations</i>	<i>XIII</i>
<i>Introduction</i>	1
<i>Aim of the work</i>	29
<i>Review of literature</i>	
<i>Anatomical and histological aspects of the liver</i>	30
<i>Budd-chiari syndrome</i>	43
• <i>Definition</i>	19
• <i>Classification</i>	20
• <i>Epidemiology</i>	23
• <i>Pathology of BCS</i>	48
• <i>Etiology of BCS</i>	52
• <i>Clinical manifestations of BCS</i>	84
• <i>Diagnosis of BCS</i>	90
<i>Management strategies, prognosis and survival of BCS</i>	128
<i>Patients and methods</i>	167
<i>Results</i>	181
<i>Illustrative figures</i>	233
<i>Discussion</i>	240
<i>Summary</i>	263
<i>Conclusions and recommendations</i>	270
<i>References</i>	272
<i>Arabic summary</i>	

LIST OF TABLES

Tab. No.	Title	Page No.
<u>Table (1):</u>	Causes of BCS.....	54
<u>Table (2):</u>	Sapporo classification criteria for the Antiphospholipid Syndrome.....	68
<u>Table (3):</u>	French-American-British (FAB) and World Health Organization (WHO) classification of MPDs	71
<u>Table (4):</u>	Warfarin properties	131
<u>Table (5):</u>	Unfractionated heparin properties	133
<u>Table (6):</u>	Low molecular weight heparin (LMWH) properties.....	134
<u>Table (7):</u>	Indications for TIPS.....	148
<u>Table (8):</u>	Contraindications for Placement of TIPS	149
<u>Table (9):</u>	(Rotterdam) Murad's Budd-Chiari syndrome prognostic classification.....	164
<u>Table (10):</u>	Grade of esophageal varices (Three- size classification.....	171
<u>Table (11):</u>	Baveno scoring system for portal hyper- tensive gastropathy (PHG)	171
<u>Table (12):</u>	WHO performance status scale	177
<u>Table (13):</u>	The schedule of post-intervention follow up.....	179
<u>Table (14):</u>	Age and Gender distribution among the studied patients	181
<u>Table (15):</u>	Residence distribution among the studied patients	182
<u>Table (16):</u>	Classification of patients according to the onset of the disease	183



LIST OF TABLES (Cont...)

Tab. No.	Title	Page No.
<u>Table (17):</u>	<u>Child-Pugh score classification of patients.....</u>	<u>184</u>
<u>Table (18):</u>	<u>WHO performance status classification of patients</u>	<u>185</u>
<u>Table (19):</u>	<u>Frequency of inherited procoagulative disorders among studied patients.....</u>	<u>187</u>
<u>Table (20):</u>	<u>Frequency of different acquired procoagulative disorders.....</u>	<u>188</u>
<u>Table (21):</u>	<u>Frequency of use of different types of hormonal therapy in females</u>	<u>189</u>
<u>Table (22):</u>	<u>Status of etiologies of BCS (combined, single, idiopathic) in the studied cases</u>	<u>190</u>
<u>Table (23):</u>	<u>Frequency of different symptoms and signs in the studied patients</u>	<u>191</u>
<u>Table (24):</u>	<u>Baseline radiological criteria of the studied patients</u>	<u>194</u>
<u>Table (25):</u>	<u>Pattern of the occlusion of hepatic veins in studied patients.....</u>	<u>195</u>
<u>Table (26):</u>	<u>Baseline relevant endoscopic findings of the studied patients.....</u>	<u>197</u>
<u>Table (27):</u>	<u>Baseline results of ascetic sampling of the studied patients</u>	<u>198</u>
<u>Table (28):</u>	<u>Baseline laboratory data of the studied patients</u>	<u>199</u>
<u>Table (29):</u>	<u>Main indication for intervention in the studied patients</u>	<u>200</u>

LIST OF TABLES (Cont...)

Tab. No.	Title	Page No.
Table (30):	<u>Site of conducted Intervention (Angioplasty with Stenting) in the studied patients</u>	<u>201</u>
Table (31):	<u>Operative and early postoperative (1 - week) complications in the studied cases.....</u>	<u>202</u>
Table (32):	<u>One year stent patency</u>	<u>205</u>
Table (33):	<u>Stent occlusion during and at the end of follow up.....</u>	<u>205</u>
Table (34):	<u>Abdominal pain before intervention, during and at the end of follow up (at 1, 3, 6, 12 months).....</u>	<u>206</u>
Table (35):	<u>Frequency of abdominal enlargement before intervention, during and at the end of follow up.....</u>	<u>207</u>
Table (36):	<u>Status of ascites (as detected clinically and by US) before intervention, during and at the end of follow up.....</u>	<u>208</u>
Table (37):	<u>Hepatic tenderness before intervention, during and at the end of follow up.....</u>	<u>209</u>
Table (38):	<u>Dilated veins on the abdomen before intervention, during and at the end of follow up.....</u>	<u>210</u>
Table (39):	<u>Comparison between child class before and after intervention in the studied patients</u>	<u>211</u>

LIST OF TABLES (Cont...)

Tab. No.	Title	Page No.
Table (40):	Comparison between WHO performance status before and after intervention in the studied patients.....	212
Table (41):	Comparison between endoscopic finding before and after intervention in the studied patients	213
Table (42):	Laboratory investigations before intervention, during and at the end of follow up (at 1, 3, 6, 12 months):	215
Table (43):	Mean size of the liver (cm) before and after intervention.....	217
Table (44):	Mean size of the spleen (cm) before and after intervention.....	218
Table (45):	Mean Portal vein diameter (mm) before intervention, and at the end of follow up	219
Table (46):	Portal vein flow direction before intervention, and at the end of follow up.....	221
Table (47):	Portal vein velocity (mm/sec) before intervention and at the end of follow up.....	223

LIST OF TABLES (Cont...)

Tab. No.	Title	Page No.
<u>Table (48):</u>	<u>One year post intervention survival among patients.....</u>	<u>232</u>
<u>Table (49):</u>	<u>The etiology of BCS in different studies compared to the current study</u>	<u>249</u>
<u>Table (50):</u>	<u>Clinical manifestations of BCS in different studies compared to the current study.....</u>	<u>251</u>
<u>Table (51):</u>	<u>Outcome of PTBA with stenting in BCS patients in different studies compared to the current study.....</u>	<u>260</u>

LIST OF FIGURES

Fig. No.	Title	Page No.
<u>Fig. (1):</u>	Segmental anatomy of the liver.....	31
<u>Fig. (2):</u>	Vascular anatomy of the liver.....	32
<u>Fig. (3):</u>	Represent the portal veins.....	34
<u>Fig. (4):</u>	Diagram of the distribution of the four main portal veins to the segments of the liver and the hepatic venous drainage to the inferior vena cava.....	37
<u>Fig. (5):</u>	Inferior vena cava	38
<u>Fig. (6):</u>	Microvascular anatomy of the liver	40
<u>Fig. (7):</u>	Zones of the liver parenchyma.....	42
<u>Fig. (8):</u>	Histological examination shows sinusoidal dilatation and congestion	48
<u>Fig. (9):</u>	The protein C/protein S pathway.....	61
<u>Fig. (10):</u>	Anticoagulant activities of Antithrombin	62
<u>Fig. (11):</u>	Causes of Hyperhomocysteinemia	65
<u>Fig. (12):</u>	Proposed algorithm for a diagnosis of myeloproliferative disease (MPD) in patients with primary Budd-Chiari syndrome	75
<u>Fig. (13):</u>	Normal triphasic hepatic vein flow.....	95
<u>Fig. (14):</u>	Thrombosis of the inferior vena cava on B-mode US	97
<u>Fig. (15):</u>	Inferior right hepatic vein	99
<u>Fig. (16):</u>	Dilated caudate veins	99

LIST OF FIGURES (Cont...)

Fig. No.	Title	Page No.
<u>Fig. (17):</u>	Intrahepatic collateral veins suggestive of Budd-Chiari syndrome.....	<u>100</u>
<u>Fig. (18):</u>	Flow disturbances in Budd-Chiari syndrome.....	<u>101</u>
<u>Fig. (19):</u>	Occlusion of upper part of inferior vena cava (IVC)	<u>102</u>
<u>Fig. (20):</u>	Hypertrophy of caudate lobe	<u>103</u>
<u>Fig. (21):</u>	Inhomogenous enhancement of liver	<u>106</u>
<u>Fig. (22):</u>	Parenchymal changes in Budd-Chiari syndrome.....	<u>106</u>
<u>Fig. (23):</u>	CT in a patient with acute type Budd-Chiari syndrome secondary to thrombosis of the hepatic veins.....	<u>108</u>
<u>Fig. (24):</u>	CT of the abdomen demonstrates lack of opacification of the hepatic veins, ascites and patchy enhancement of the porta hepatis in a patient with acute Budd-Chiari syndrome	<u>108</u>
<u>Fig. (25):</u>	CT in a 28-year-old female with idiopathic subacute Budd-Chiari syndrome.....	<u>109</u>
<u>Fig. (26):</u>	Subacute Budd-Chiari syndrome	<u>110</u>
<u>Fig. (27):</u>	CT in a patient with chronic Budd-Chiari syndrome	<u>110</u>
<u>Fig. (28):</u>	CT in a patient with chronic Budd-Chiari syndrome	<u>111</u>

LIST OF FIGURES (Cont...)

Fig. No.	Title	Page No.
<u>Fig. (29):</u>	A 42-year-old female patient with chronic Budd-Chiari syndrome demonstrates large enhancing regenerative nodule	112
<u>Fig. (30):</u>	MR angiography shows benign regenerative nodules.....	114
<u>Fig. (31):</u>	Hepatocellular carcinoma (HCC) in Budd-Chiari syndrome	115
<u>Fig. (32):</u>	Hypoperfusion in peripheral portion of liver	116
<u>Fig. (33):</u>	Caudate lobe hypertrophy	117
<u>Fig. (34):</u>	Short segment obstruction in inferior vena cava.....	119
<u>Fig. (35):</u>	Stenosis of inferior vena cava (IVC) due to external compression of enlarged caudate lobe.	119
<u>Fig. (36):</u>	Intrahepatic collateral veins.....	120
<u>Fig. (37):</u>	Collateral veins in Budd-Chiari syndrome	121
<u>Fig. (38):</u>	Proposed diagnostic strategy for Budd-Chiari syndrome	126
<u>Fig. (39):</u>	Percutaneous transhepatic balloon angioplasty was performed and the right hepatic vein was widely patent after balloon angioplasty.....	143
<u>Fig. (40):</u>	Percutaneous angioplasty of segmental stenosis of inferior vena cava	146

LIST OF FIGURES (Cont...)

Fig. No.	Title	Page No.
<u>Fig. (41):</u>	A Covered stent TIPS occlusion with symptomatic recurrence of ascites. B Restoration of flow in TIPS after balloon dilatation	<u>151</u>
<u>Fig. (42):</u>	Mesocaval shunt	<u>152</u>
<u>Fig. (43):</u>	Contrast-enhanced transverse CT in a patient with chronic Budd-Chiari syndrome demonstrates multiple regenerative nodules (small arrows) as well as contrast enhancement of a mesocaval shunt indicating patency	<u>156</u>
<u>Fig. (44):</u>	Mesoatrial shunt	<u>156</u>
<u>Fig. (45):</u>	Recommended treatment strategy in patients with primary Budd-Chiari syndrome	<u>163</u>
<u>Fig. (46):</u>	Gender distribution among the studied patients	<u>181</u>
<u>Fig. (47):</u>	Residence distribution among the studied patients	<u>182</u>
<u>Fig. (48):</u>	Classification of patients according to the onset of the disease.....	<u>183</u>
<u>Fig. (49):</u>	Child-Pugh score classification of patients.....	<u>184</u>
<u>Fig. (50):</u>	WHO performance status classification of patients.....	<u>186</u>

LIST OF FIGURES (Cont...)

Fig. No.	Title	Page No.
<u>Fig. (51):</u>	<u>Frequency of inherited procoagulative disorders among studied patients.....</u>	<u>187</u>
<u>Fig. (52):</u>	<u>Frequency of different acquired procoagulative disorders.....</u>	<u>188</u>
<u>Fig. (53):</u>	<u>Frequency of use of different types of hormonal therapy in females.....</u>	<u>189</u>
<u>Fig. (54):</u>	<u>Status of etiologies of BCS (combined, single, idiopathic) in the studied cases.....</u>	<u>190</u>
<u>Fig. (55):</u>	<u>Frequency of different symptoms in studied patients.....</u>	<u>192</u>
<u>Fig. (56):</u>	<u>Frequency of different signs in studied patients.....</u>	<u>193</u>
<u>Fig. (57):</u>	<u>Baseline radiological findings of the studied patients.....</u>	<u>196</u>
<u>Fig. (58):</u>	<u>Baseline endoscopic criteria of the studied patients.....</u>	<u>197</u>
<u>Fig. (59):</u>	<u>Results of total protein in ascitic fluid and serum ascites albumin gradient (SAAG).....</u>	<u>198</u>
<u>Fig. (60):</u>	<u>Main indication for intervention in the studied patients.....</u>	<u>200</u>
<u>Fig. (61):</u>	<u>Type of Intervention done in the studied patients.....</u>	<u>201</u>
<u>Fig. (62):</u>	<u>Procedure complications.....</u>	<u>202</u>
<u>Fig. (63):</u>	<u>One year stent patency</u>	<u>205</u>

X