Outcome of Transjugular Intrahepatic Portosystemic Shunt (TIPS) in Budd-Chiari Syndrome patients Single centre experience

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List of Abbreviations

ALP	Alkaline phosphatase
APAs	Antiphospholipid antibodies
APC	Activated protein C
APLAS	Antiphospholipid antibody syndrome
APTT	Activated partial thromboplastin time
AT III	Antithrombin III
BCS	Budd-Chiari syndrome
BCSG	Budd-Chiari Study Group
CT	Computed Tomography
DIC	Disseminated intravascular coagulation
DVT	Deep venous thrombosis
FVLM	Factor V Leiden mutation
HH	Hyperhomocysteinemia
HVOO	Hepatic venous outflow obstruction
HVPG	Hepatic venous pressure gradient
HVs	Hepatic veins
IVC	Inferior vena cava
JAK_2	Janus tyrosine kinase-2
LA	Lupus Anticoagulant
LAP	Leukocyte alkaline phosphatase
LHV	Left hepatic vein
LMWH	Low molecular weight heparin
MELD	Model for end-stage liver disease
MHV	Middle hepatic vein
MPDs	Myeloproliferative disorders

List of Abbreviations *(Cont.)*

MRI	Magnetic resonance imaging
MRV	Magnetic resonance venography
MSCT	Multislice CT
MTHFR	Methylenetetrahydro-folate reductase
OCPs	Oral Contraceptive Pills
P. ANCA	Peripheral anti neutrophylic cytoplasmic antibody
PC	Protein C
PGM	Prothrombin gene mutation
PHG	Portal hypertensive gastropathy
PI	Prognostic index
PM	Prothrombin mutation
PNH	Paroxysmal nocturnal hemoglobinuria
PPG	Portal pressure gradient
PS	Protein S
PTFE	Polytetrafluoroethylene
PV	Portal vein
PVT	Portal vein thrombosis
RHV	Right hepatic vein
rTPA	Recombinant tissue plasminogen activator
TIPS	Transjugular Intrahepatic Portosystemic Shunt
UNOS	United Network for Organ Sharing
US	Ultrasound

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Introduction

Budd-Chiari syndrome is a congestive hepatopathy caused by obstruction of the hepatic venous outflow at any level from the small hepatic veins to the junction of the inferior vena cava (IVC) with the right atrium ***(Valla, 2009).***

George Budd described this hepatic disorder in 1845, and **Chiari** added the first pathologic description of a liver with "obliterating endophlebitis of the hepatic veins" in 1899 ***(Chung et al., 2006).***

The hepatic outflow obstruction results in portal hypertension and sinusoidal congestion with ischaemic hepatocyte dysfunction.

This causes the dominant clinical features of abdominal pain, hepato- and splenomegaly, ascites, and oesophageal varices and the development of fulminant hepatic failure ***(Valla, 2009).***

The most common underlying disorders in patients with BCS are myeloproliferative disorders such as polycythaemia vera and essential thrombocytosis. Some patients may have a combination of thrombophilias ***(Ranjani et al., 2008).***

Other common associations include oral contraceptive use, protein C and S deficiencies, antiphospholipid syndrome, antithrombin III deficiency, paroxysmal nocturnal

haemoglobinuria, malignancy, Behcet's syndrome and trauma ***(Denninger et al., 2000).***

The Budd–Chiari syndrome is termed primary or secondary based on the aetiology and site of venous outflow obstruction. Primary BCS originates from within the lumen of the veins or venules and results from thrombosis, webs or endophlebitis. Secondary BCS results from an extra-luminal lesion such as tumour, abscess or cyst, which can invade the lumen or cause extrinsic compression of the hepatic venous outflow tract ***(Hoekstra and Janssen, 2008).***

Four main clinical variations have been described: fulminant, acute, subacute or chronic ***(Darwish et al., 2009).***

The subacute form is the most common presentation and is characterized by slow accumulation of ascites. Hepatocellular damage is minimal due to the development of hepatic and portal venous collaterals.

The fulminant form is rare and is defined as the development of hepatic encephalopathy within 8 weeks of the onset of jaundice.

Acute BCS has a rapid onset, intractable ascites and a short duration of symptoms. Collateral venous channels do not develop in the acute form of BCS. Chronic BCS usually occurs in the background of cirrhosis.

Early diagnosis is fundamental, Doppler-ultrasound, computed tomography or magnetic resonance imaging of hepatic veins and inferior vena cava are usually successful in demonstrating non-invasively the obstacle or its consequences, the collaterals to hepatic veins or inferior vena cava

Treatment depends on the underlying cause, the anatomic location, the extent of the thrombotic process and the severity of liver disease and it can be medical, surgical, or interventional. A therapeutic strategy has been proposed where anticoagulation, correction of risk factors, diuretics and prophylaxis for portal hypertension are used first; then angioplasty for short-length venous stenosis or TIPS for long segment occlusion, and ultimately liver transplantation ***(Valla, 2009)***.

Interventional radiology options as angioplasty and transjugular intrahepatic portosystemic shunt (TIPS) should be considered. This may reverse the hepatic congestion and halt the progression of the disease ***(Li et al., 2009)***.

TIPS decompress the liver by creating an alternative venous outflow tract ***(Panagiotou et al., 2007).*** It is particularly useful, either alone or as a bridge to liver transplantation, in patients with an acute presentation such as those with variceal bleeding, and patients with fulminant

hepatic failure or chronic illness in whom thrombolysis and angioplasty were unsuccessful ***(Khuroo et al., 2005).***

TIPS may be preferred over surgical shunting because it avoids laparotomy and has less periprocedure mortality and morbidity ***(Corso et al., 2008).*** Although long-term patency rate is only about 49%, patients in whom stent stenosis occurs do not generally worsen, perhaps because the shunt allows time for collateral circulation to develop ***(Perello et al., 2002).***

The recently introduced, polytetrafluoroethylene-covered stents may improve the patency rates and have been shown to reduce the incidence of TIPS dysfunction in BCS patients ***(Cura et al., 2008 and Murad et al., 2008).*** Other important complications of TIPS procedure is the occurrence of thrombosis, infection and portosystemic encephalopathy ***(Eldorry et al., 2011)***.

Aim of the work

The aim of this study is to:

1- Determine the short term outcome of TIPS in Budd-Chiari syndrome patients in the form of immediate post procedure complications and possible effect of procedure on presenting symptoms.

2- Determine Long term outcome of TIPS in the form of stent patency and patient survival.